European Paediatric Ophthalmological Society
39th Annual Meeting

Programme
and
Book of Abstracts
European Paediatric Ophthalmological Society

EPOS 2013

39th Annual Meeting

Programme
and
Book of Abstracts

Leiden, the Netherlands
October 11-12, 2013

http://epos-focus.org/meeting/2013
meeting@epos-focus.org
We would like to thank our sponsors and exhibitors

**Gold sponsor**

Clarity Medical Systems

**Silver sponsors**

<table>
<thead>
<tr>
<th>Abbott Medical Optics</th>
<th>Théa Pharma</th>
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<tr>
<td><img src="image1.png" alt="Abbott Medical Optics" /></td>
<td><img src="image2.png" alt="Théa Pharma" /></td>
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<td>Bausch + Lomb</td>
<td>I2eyediagnostics</td>
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<td><img src="image4.png" alt="I2eyediagnostics" /></td>
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**Other sponsors and exhibitors**

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<tr>
<th>Optiplus</th>
<th>BcnInnova</th>
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<tr>
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<td>Metrovision</td>
<td>Eastbeco Trusetal Verbandstoffwerke GmbH</td>
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<td><img src="image10.png" alt="Eastbeco Trusetal Verbandstoffwerke GmbH" /></td>
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</table>
Dear colleagues and friends,

A warm welcome to Leiden! As president and local organizer I hope you will enjoy this 39th annual meeting of the European Paediatric Ophthalmological Society EPOS. Leiden offers a lot of possibilities. It accommodates the oldest university in the Netherlands, founded in 1575 by William, Prince of Orange, who was the leader of the Dutch Revolt in the Eighty Years’ War. Maybe you will have the opportunity to walk around and enjoy.

At the moment I write this introduction a record number of 248 participants from 41 countries have registered. This offers you the opportunity to share your thoughts with a diverse audience. The main theme of this years’ meeting is paediatric ophthalmic tumors with a side step to limbal stemcell insufficiency. The organizing committee intended to compose a programme of diversity and high scientific level. We hope that in the end, you will be able to look back at a fruitful meeting.

The number of applications increases every year, so the scientific committee could not award an oral presentation to everybody. We therefore decided to introduce rapid fire presentations in combination with posters and make posters easily accessible. Authors of posters are requested to attend their work when scheduled. As always, best paper and poster presentations for presenters of 35 years or younger will be awarded at the end of the meeting. For this, the abstract as well as the presentation of the paper or poster will be taken into account.

A new development in our field is the initiative of several societies to join forces and collaborate. We will discuss this in detail at the general assembly on Friday afternoon. Here Nikolas Ziakas will take over as president of the Society and two new board members will be elected. If you are a member of EPOS, please come to this meeting and share with us your thoughts about the future of our Society.

Afterwards we will join for the more relaxing part of the program. At the National Museum of Antiquities under the watchful eye of the Sphinx you will be able to reminisce, exchange ideas and walk around the exposition.

Several exhibitors were willing to support this meeting. I would like to express our sincere gratitude for their significant efforts and encourage all participants to visit the technical exhibit during breaks.

This year we celebrate the 300 anniversary of the Treaty of Utrecht, a series of individual peace treaties between several European states. Even then, people realized that cooperation is indispensable for progress. I hope this meeting will inspire you, will initiate new collaborations and leaves you with many new friends!

After 3 years I hand over the presidency to Nikolas Ziakas. I wish him a lot of inspiration. EPOS is a wonderful society. Not only do we learn much of each other, we are also willing to spread knowledge about paediatric ophthalmology around the world. In my opinion a relative small society like ours can only flourish with the help of many dedicated professionnals. So if you have ideas about how EPOS should perform or move forward, do not linger in the corridors but step forward and contribute.

Thank you very much for the opportunity to be your president.

Nicoline Schalij-Delfos, president
Please note that from the LUMC, you can also walk through the train station to go to the city center.

- **A** Congress Site, Building 3, Leiden University Medical Center, Hippocratespad 21
- **B** LUMC, Building 1
- **C** Reception and drinks: Faculty Club, Rapenburg 73
- **D** Dinner location: Rijksmuseum van Oudheden, Rapenburg 28 (Dutch National Museum of Antiquities)
- **E** Golden Tulip Hotel, Schipholweg 3
Registration for EPOS 2013

Thursday October 10, 2013:

18.00 – 21.30 Congress Bag Counter opens for registration of pre-registered participants and onsite registration in Leiden University Medical Center (LUMC), building 3

19.30 Opening buffet for all participants at Congress site

LUMC, Foyer building 3

LEGEND

1. Entrance
2. Registration desk
3. Wardrobe
4. Elevators
5. Lockers
6. Staircase
7. Coffee
8. Coat rack
9. ~ 18 Booths

Friday Oct 11
Foyer, Posters A no. 1-16
Foyer, Posters B no. 17-28
1st floor Posters C, no. 29-37

Saturday Oct 12
Foyer, Posters A no. 38-53
Foyer, Posters B no. 54-66

GROUND FLOOR
General notifications

Presentations: participants are advised to upload their presentation well in advance. It will not be possible to use a personal laptop. You can upload your presentation in the preview room V1-22. Speakers of the first session on Friday can upload their presentation on Thursday evening at the registration desk.

Presentation time will be 7 minutes for free papers and 2 minutes for rapid fire (RF) presentations. Time for discussion will be 3 minutes for free papers and 1 minute for RF presentations. You are kindly requested not to exceed the assigned time.

For the RF presentations 2 lecterns will be available. You are requested to take your position at the other lectern during the 1 minute questioning of your predecessor.

Posters will be displayed for 1 day: posters 1–37 on Friday and 38–66 on Saturday. You will find the pre-numbered poster boards in the lobby and on the first floor.

Grants: at the end of the meeting grants for best presentation (free paper and rapid fire) and poster will be awarded. Any registered member participating in the annual meeting aged 35 years or less may apply for this award which will be decided upon by the Board of the Society. The actual presentation as well as the response to questions about the paper or poster will be taken into account.

For assistance please contact the registration desk. Contact number only during the meeting for urgent matters: +31-71-526 7976.

Accreditation: The 39th Annual Meeting of the European Paediatric Ophthalmological Society (Leiden, the Netherlands, 11.–12.10.2013) has been granted 12 European CME credits (ECMEC) by the European Accreditation Council for Continuing Medical Education (EACCME). Event code 9152.

The Netherlands Ophthalmological Society (NOG) and the Netherlands Society for Orthoptics (NVvO) have granted 12 accreditation points.

Reception and dinner on Friday night are on 10 minutes walking distance from the Congress location.
### Scientific Programme EPOS, October 11-12, 2013

#### Friday October 11, 2013:

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
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<tbody>
<tr>
<td>8.30</td>
<td>Welcome President EPOS / local organizer Nicoline Schalij-Delfos</td>
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<tr>
<td>8.35</td>
<td>Welcome Prof Dr J.B.M.Z. Trimbos, Chair 3rd Clinical Division LUMC</td>
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<tr>
<td>8.45</td>
<td><strong>Gliomas</strong></td>
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<tr>
<td>8.45 – 9.05</td>
<td>L1  Neurosurgery in Optic Pathway Gliomas</td>
</tr>
<tr>
<td>9.05 – 9.25</td>
<td>L2  Chemotherapy in NF and tumor vaccination</td>
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<tr>
<td>9.25 – 9.35</td>
<td>Discussion</td>
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<tr>
<td>9.35 – 9.45</td>
<td>FP1  The ophthalmological examination in neurofibromatosis type 1</td>
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<tr>
<td>9.45 – 9.55</td>
<td>FP2  Optic Pathway Gliomas: Which role plays the ophthalmologist?</td>
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<tr>
<td>9.55 – 10.05</td>
<td>FP3  Tele-ROP for middle-income countries: 5 years of 'KIDROP' in India and the roadmap for universal infant eye screening</td>
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<tr>
<td>10.05 – 10.15</td>
<td>FP4  The first results of the implementation of screening and treatment ROP in Astana</td>
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<tr>
<td>10.15 – 10.25</td>
<td>FP5  Visual functions at 2.5 years in two cohorts of prematurely-born children in Sweden</td>
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<tr>
<td>10.25 – 10.55</td>
<td>Coffee break / Poster exhibition P01 – P13</td>
</tr>
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</table>

#### Moderator: Branca Stirn Kranjc

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
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<tbody>
<tr>
<td>10.55 – 11.10</td>
<td>L3  Neurofibromatosis type II in childhood</td>
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<tr>
<td>11.10 – 11.30</td>
<td>L4  Screening for optic pathway gliomas</td>
</tr>
<tr>
<td>11.30 – 11.45</td>
<td>Round Table: Screening for NF type 1 Panel: Ian Simmons, Sofie Caen, Branca Stirn Kranjc</td>
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<tr>
<td>Time</td>
<td>RF</td>
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<td>11.45 – 11.48</td>
<td>RF01</td>
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<td>11.48 – 11.51</td>
<td>RF02</td>
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<td>11.51 – 11.53</td>
<td>RF03</td>
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<td>11.53 – 11.56</td>
<td>RF04</td>
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<td>11.56 – 11.59</td>
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<tr>
<td>11.59 – 12.02</td>
<td>RF06</td>
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<tr>
<td>12.02 – 12.05</td>
<td>RF07</td>
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<td>12.05 – 12.08</td>
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<td>12.08 – 12.11</td>
<td>RF09</td>
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<td>12.11 – 12.14</td>
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<td>12.14 – 12.17</td>
<td>RF11</td>
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<td>12.20 – 12.25</td>
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<tr>
<td>12.25 – 12.45</td>
<td>L5</td>
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<td>12.45 – 13.45</td>
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<td>13.45 – 14.00</td>
<td>L6</td>
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<td>14.00 – 14.05</td>
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<tr>
<td>Time</td>
<td>Session</td>
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<tr>
<td>14.05 – 14.15</td>
<td>FP6</td>
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<td>14.15 – 14.25</td>
<td>FP7</td>
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<td>14.25 – 14.35</td>
<td>FP8</td>
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<tr>
<td>14.45 – 14.48</td>
<td>RF12</td>
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<tr>
<td>14.48 – 14.51</td>
<td>RF13</td>
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<td>14.51 – 14.54</td>
<td>RF14</td>
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<tr>
<td>14.54 – 14.57</td>
<td>RF15</td>
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<tr>
<td>14.57 – 15.00</td>
<td>RF16</td>
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<tr>
<td>15.00 – 15.03</td>
<td>RF17</td>
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<td>15.05 – 15.30</td>
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<tr>
<td>15.30 – 15.50</td>
<td>L7</td>
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<tr>
<td>15.50 – 16.00</td>
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<td>16.00 – 17.15</td>
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SOCIAL PROGRAM

Friday October 11, 2013:

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
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<tbody>
<tr>
<td>18.30</td>
<td>Reception Faculty Club, Rapenburg 73*</td>
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<tr>
<td>19.30 – 23.00</td>
<td>Dinner in the Temple Hall of the Museum of Antiquities (Rijksmuseum van Oudheden), Rapenburg 28*</td>
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</tbody>
</table>

* See map on page 8.
**Saturday October 12, 2013:**

<table>
<thead>
<tr>
<th>Time</th>
<th>Session</th>
<th>Topic</th>
<th>Presenter</th>
<th>Country</th>
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</thead>
<tbody>
<tr>
<td>8.30</td>
<td>L8</td>
<td>An introduction to new and future treatment approaches to intraocular retinoblastoma</td>
<td>A Linn Murphree</td>
<td>USA</td>
</tr>
<tr>
<td>8.30 – 8.55</td>
<td>L9</td>
<td>Imaging of paediatric ophthalmic tumors: towards new guidelines for MR imaging</td>
<td>Pim de Graaf</td>
<td>Netherlands</td>
</tr>
<tr>
<td>9.15</td>
<td></td>
<td>Discussion</td>
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<tr>
<td>9.15 – 9.25</td>
<td>FP10</td>
<td>The uptake of prenatal testing for retinoblastoma susceptibility by pregnant women at increased risk</td>
<td>Charlotte Dommering</td>
<td>Netherlands</td>
</tr>
<tr>
<td>9.25 – 9.35</td>
<td>FP11</td>
<td>Retinal detachment after retinoblastoma treatment</td>
<td>Pascal Dureau</td>
<td>France</td>
</tr>
<tr>
<td>9.35 – 9.45</td>
<td>FP12</td>
<td>Evaluating the risk of extraocular tumor spread following intravitreal injection therapy for retinoblastoma: a systematic review</td>
<td>Stephen Smith</td>
<td>USA</td>
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<tr>
<td>9.45 – 9.55</td>
<td>FP13</td>
<td>Outcomes for limbal cataract aspiration and IOL implantation in children with radiation-induced cataracts in retinoblastoma</td>
<td>Marta Morales</td>
<td>Spain</td>
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<tr>
<td>10.05 – 10.25</td>
<td>L10</td>
<td>Quiz: Differential diagnosis of ‘white retinal tumors’</td>
<td>Annette Moll</td>
<td>Netherlands</td>
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<td>10.25 – 10.55</td>
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<td>Coffee break / Poster exhibition P38 – P50</td>
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<td></td>
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<td><strong>New treatment strategies</strong></td>
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<td><strong>Moderators: A Linn Murphree / Annette Moll</strong></td>
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<tr>
<td>11.05 – 11.20</td>
<td>L11</td>
<td>Intra-arterial treatment of retinoblastoma</td>
<td>Machteld Bosscha</td>
<td>Netherlands</td>
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<tr>
<td>11.20 – 11.35</td>
<td>L12</td>
<td>Intra-vitreal treatment of retinoblastoma</td>
<td>Francis Munier</td>
<td>Switzerland</td>
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<tr>
<td>11.35 – 11.50</td>
<td>L13</td>
<td>Genetic and epigenetic changes in retinoblastoma</td>
<td>Dietmar Lohmann</td>
<td>Germany</td>
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<td>11.50 – 12.00</td>
<td>FP14</td>
<td>Direct intra-arterial (ophthalmic artery) chemotherapy for advanced intraocular retinoblastoma: 5 years’ experience</td>
<td>Doris Hadjistilianou</td>
<td>Italy</td>
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<tr>
<td>Time</td>
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<tr>
<td>12.00 – 12.03</td>
<td>RF18</td>
<td>Intravitreal Chemotherapy – the first line of Retinoblastoma salvage eye treatment</td>
<td>Nadiya Bobrova</td>
<td>Ukraine</td>
</tr>
<tr>
<td>12.03 – 12.06</td>
<td>RF19</td>
<td>The response of retinoblastoma to local treatment in chemotherapy lack of effectiveness</td>
<td>Evgeniya Bulgakova</td>
<td>Russia</td>
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<tr>
<td>12.06 – 12.09</td>
<td>RF20</td>
<td>Macular retinoblastoma – An outcome study following primary chemotherapy alone</td>
<td>John Bladen</td>
<td>UK</td>
</tr>
<tr>
<td>12.09 – 12.12</td>
<td>RF21</td>
<td>Macular retinoblastoma &amp; visual function: beyond visual acuity</td>
<td>Pablo Durán-Pérez</td>
<td>Spain</td>
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<tr>
<td>12.12 – 12.15</td>
<td>RF22</td>
<td>Choroidal osteoma: rare familial occurrence</td>
<td>Mary van Schooneveld</td>
<td>Netherlands</td>
</tr>
<tr>
<td>12.15 – 12.18</td>
<td>RF23</td>
<td>Pathological analysis of pediatric ocular tumors</td>
<td>Mària Csídey</td>
<td>Hungary</td>
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<tr>
<td>12.20 – 12.40</td>
<td></td>
<td>Round Table: Treatment for retinoblastoma</td>
<td><strong>Moderator: A Linn Murphree</strong></td>
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<td>Panel: Machteld Bosscha, Francis Munier, Dietmar Lohmann, Doris Hadjistilianou, Annette Moll</td>
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<tr>
<td>12.40 – 12.50</td>
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<td>Discussion</td>
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<tr>
<td>12.50 – 13.50</td>
<td></td>
<td>Lunch / Poster exhibition P51 – P67</td>
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<tr>
<td>13.50</td>
<td></td>
<td><strong>Orbital and vascular lesions</strong></td>
<td><strong>Moderators: Nikolas Ziakas / Gré Luyten</strong></td>
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<tr>
<td>13.50 – 14.15</td>
<td>L14</td>
<td>Benign periocular masses in childhood</td>
<td>Geoffrey Rose</td>
<td>UK</td>
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<tr>
<td>14.15 – 14.20</td>
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<td>Discussion</td>
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<tr>
<td>14.20 – 14.30</td>
<td>FP15</td>
<td>Suspected optic disc oedema in children – how helpful is ultrasound ONSD measurement in clinical decision making?</td>
<td>Anne Cees Houtman</td>
<td>Netherlands</td>
</tr>
<tr>
<td>14.30 – 14.40</td>
<td>FP16</td>
<td>Histiocytic lesion of the orbit in children</td>
<td>Joan Prat</td>
<td>Spain</td>
</tr>
<tr>
<td>14.40 – 14.50</td>
<td>FP17</td>
<td>Morbus Graves in children and adolescents</td>
<td>Irene Notting</td>
<td>Netherlands</td>
</tr>
<tr>
<td>14.50 – 15.00</td>
<td>FP18</td>
<td>Melanocytic tumors and their look alikes in and around the eye</td>
<td>Evelyne van de Winkel</td>
<td>Netherlands</td>
</tr>
<tr>
<td>15.00 – 15.10</td>
<td>FP19</td>
<td>Clinical features, instrumental diagnostics and surgical treatment of orbital rhabdomyosarcoma in children</td>
<td>Svetlana Saakyan</td>
<td>Russia</td>
</tr>
<tr>
<td>Time</td>
<td>Session</td>
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<tr>
<td>15.15 – 15.30</td>
<td>L15</td>
<td>Towards reliable VEGF concentration measurements in blood samples of infants and young children</td>
<td>Birgit Lorenz</td>
<td>Germany</td>
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<td>15.30 – 15.40</td>
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<td>Discussion</td>
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<td>15.40 – 16.10</td>
<td></td>
<td>Tea break</td>
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<tr>
<td><strong>Moderators:</strong> Chris Lloyd / Martha Tjon Fo San</td>
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<tr>
<td>16.10 – 16.13</td>
<td>RF24 (P57)</td>
<td>Congenital nasolacrimal duct obstruction: a definitive approach to management of a common condition</td>
<td>Maria Napier</td>
<td>UK</td>
</tr>
<tr>
<td>16.13 – 16.16</td>
<td>RF25 (P58)</td>
<td>An unusual evolution of preseptal cellulitis to subperiosteal abscess</td>
<td>Athanasios Fitsios</td>
<td>Greece</td>
</tr>
<tr>
<td>16.19 – 16.22</td>
<td>RF27 (P52)</td>
<td>Deep periocular infantile capillary haemangiomas responding to topical timolol maleate</td>
<td>Kanmin Xue</td>
<td>UK</td>
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<tr>
<td>16.22 – 16.25</td>
<td>RF28 (P53)</td>
<td>Active surgical management in infantile capillary hemangiomas of the orbit and eyelids</td>
<td>Svitlana Tronina</td>
<td>Ukraine</td>
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<tr>
<td>16.25 – 16.28</td>
<td>RF29 (P62)</td>
<td>A clinical classification system for Paravenous Pigmentary Chorioretinal Atrophy (PPCRA) in Childhood</td>
<td>Göran Darius Hildebrand</td>
<td>UK</td>
</tr>
<tr>
<td>16.28 – 16.31</td>
<td>RF30 (P63)</td>
<td>Choroidal neovascularization secondary to Autosomal Recessive Bestrophinopathy in a 9-year-old girl treated by intravitreal ranibizumab injection</td>
<td>Marta Pawlak</td>
<td>Poland</td>
</tr>
<tr>
<td>16.31 – 16.34</td>
<td>RF31 (P64)</td>
<td>Identical Twins Exposed! – Bestrophinopathy, Autosomal Recessive?</td>
<td>Lintje Ho</td>
<td>Netherlands</td>
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<tr>
<td><strong>Moderators:</strong> Gabriela Wirth Barben / Marije Sminia</td>
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<tr>
<td>16.45 – 17.00</td>
<td>L16</td>
<td>Enucleation and the pediatric socket</td>
<td>Dyonne Hartong</td>
<td>Netherlands</td>
</tr>
<tr>
<td>17.00 – 17.20</td>
<td>L17</td>
<td>Rhabdomyosarcoma</td>
<td>Nathalie Cassoux</td>
<td>France</td>
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LECTURES
Optic Pathway Gliomas (OPG) can be distinguished into three clinical entities: optic nerve glioma, tumors of the optic nerves and chiasm and hypothalamic/chiasmatic tumors. The latter is also mentioned Hypothalamic Chiasmatic LowGrade Glioma (HCLGG), and will be focused on concerning its neurosurgical treatment aspects. HCLGG are typically bulky and exophytic tumors arising from the chiasm or along the optic tract with involvement of the hypothalamus. Clinical signs can be subtle neuroophthalmological disturbances or endocrine symptoms. Classical is the diencephalic syndrome with severe cachexia. Imaging characteristics on MR are strong enhancement with Gadolineum on T1 weighted sequences and cysts within the tumor.

The histology is always a low grade glioma. The majority of tumors are pilocytic astrocytomas(WHO grI), but a small minority of tumors have been defined as pilomyxoid astrocytomas with a less favorable course. The natural course of HCLGG is unpredictable varying form indolent to rapid progression. NeuroFibromatosis (NF) type I is strongly correlated with optic pathway gliomas in 10 – 60 % of cases mentioned in different series.

The treatment strategy in HCLGG is controversial and variable. The different treatment options are neurosurgical biopsy or resection, chemotherapy, radiation therapy and wait & scan. Many different medical doctors are involved, and a multidisciplinary pediatric neuro-oncology team is a prerequisite in order to treat and follow these patients properly. The treatment is aimed at creating a stable disease with minimal morbidity to the patient. This requires a life-long commitment of the team to the patient and the family.

Neurosurgical interventions are feasible in HCLGG patients and can contribute to creating a stable disease with minimal morbidity. However, neurosurgical interventions are always part of an individualized longterm multidisciplinary treatment strategy. Some illustrating patients will be presented and the consensus statements concerning these patients will be mentioned.

References:

Cooperative multicenter study for children and Adolescents with Low Grade Glioma. SIOP-LGG 2004
Gliomas are rare astrocytic tumors in the brain. They can present as low grade (LGG, grade I and II) or high grade (HGG, grade III and IV) gliomas. Also in the 21st century, these tumors remain challenging for the medical community. Optic pathway gliomas (OPG) in the context of NF-1 are usually grade I tumors. The major challenge is preserving maximal quality of life by avoiding tumor- and/or treatment-induced disabilities. Because of the unpredicted course of OPG, a careful assessment of the functional and radiological presentation is needed. Only in cases of visual impairment and/or radiological evidence of progression, treatment is initiated. Because of irradiation-induced side effects, particularly in children with NF-1, chemotherapy is the single option of treatment. Several first line and second line chemotherapy regimens have been implemented into clinical practice with satisfactory results. Nevertheless, optimisation of the treatment remains mandatory, and exploration of newer targeted drugs should start. A completely different situation occurs when a patient deals with HGG. The major challenge here is the improvement of overall survival, as most patients still die soon after diagnosis in spite of modern neurosurgery, radiotherapy and chemotherapy. Because of evidence of tumor-host interaction, the host can be used to obtain control over the disease via active specific immunotherapy. In this strategy, mature autologous dendritic cells are loaded with tumor antigens obtained from the autologous tumor. The innovative strategy is supported by preclinical evidence with human in vitro culture experiments and an orthotopic mouse glioma model. The technology has been transferred into clinical practice for patients with relapsed HGG, and is meanwhile also included as part of the multimodal standard therapy for patients with newly diagnosed grade IV HGG. Remarkable results motivate to further refine the technology with modern immunomodulatory approaches. Overall, two domains in neuro-oncology, OPG in NF and HGG are challenging areas. Chemotherapy in NF and tumor vaccination are beneficial in the respective diseases.
L3  Neurofibromatosis type II in childhood
Rob de Keizer
University Hospital Antwerp, Department of Ophthalmology, Antwerp, Belgium and
LUMC, Department of Ophthalmology, Leiden, the Netherlands

Purpose: To present the diagnostic steps and key-points of the monogenic autosomal
dominant NF II disease, especially the type of the severely involved adolescents with
multiple schwannomas en meningiomas.
Also to report, very shortly, the history of the introduction of the expression 'Phacomatosis'
by Prof. dr. J. vd Hoeve ophthalmologist at the Leiden University.
Material and methods: Patients were referred by one of the specialists of the LUMC skull-
base team or detected at the first visit at the orbital and neuro-ophthalmological outpatient
clinics. The well-known clinical ocular manifestations together with other rare stigmata of
NF II and their imaging of retinal and neuro-ophthalmologic tumours will be presented.
Discussion: A large variety of ocular abnormalities and deficits could be found in NF II.
Two groups could be differentiated, patients with severe involvement (with imminent
deafness and blindness) and patients who were less involved. To detect severe
disabilities in time, careful ophthalmic examinations are necessary and treatment should
be applied. Therefore screening of these infants and adolescents is important. However,
no consensus or evidence exists about time of start of screening, the kind of examinations
that should be performed and the timing of follow up examinations. Cost benefit analysis
has not been performed.
L4  Screening for optic pathway gliomas
Ian Simmons
Spire Leeds Hospital, Eye Department, Chancellor Wing, St. James University, Leeds, UK

Optic Pathway Gliomas (OPGs) are found in children both with and without neurofibromatosis type 1 (NF1). The rationale behind the creation of a screening programme will be discussed. UK paediatric ophthalmologists have the luxury of working with orthoptists who commonly deliver the majority of ophthalmic screening. The approach to screening (particularly with regard to NF1) will be presented which will highlight the patchy nature of this service and the need for a national standardised programme. ‘New’ therapies will discussed (including the use of new agents such as Iriotecan and Avastin) which offer a fresh incentive for ophthalmologists campaigning for a more organised approach to the diagnosis and management of OPGs.
Happy New Year, caustic eye trauma due to firework in Dutch children
Jan-Tjeerd de Faber
The Rotterdam Eye Hospital, Rotterdam, the Netherlands

Since 5 years Dutch Ophthalmologist monitor all firework related eye injuries during the celebrations of the New Year in the Netherlands. Each year between 250 and 350 patients are treated by ophthalmologist after being injured while using or watching firework. Consumer fire work is not allowed to be used in the Netherlands except during 16 hours around New Year. Each year on average 1.4 eyes per hour are blinded or lost to firework related injuries. Half of the victims are bystanders accidentally hit by firework ignited by other. Half of these patients are children. The Dutch experience with these injuries have led to the following combination of traumas:

- Blast injury
- Thermal trauma
- Chemical or caustic injury
- Blunt trauma
- Sharp trauma
- Skin tattoo
- Psycho-trauma.

An overview of these injuries will be presented with a pronounced advice on how to treat caustic injury in children.
L7  Limbal stem cell transplantation in childhood
Jean Jacques Gicquel
Poitier, France
As a relatively senior member of the retinoblastoma treatment fraternity, I was asked to survey the history of the management of intraocular retinoblastoma and to look into potential new modalities.

**CHEMOREDUCTION:** Primary intravenous chemotherapy (generally 6 cycles of carboplatin, etoposide and vincristine or CEV) has side effects (bone marrow suppression, low risk of hearing loss) and when used in a chemoreduction regimen with local consolidation is effective in controlling intraocular disease in slightly less than half of Group D eyes.

**INTRAERTERIAL CHEMOTHERAPY:** Regional chemotherapy in the form of intraarterial melphalan was initially demonstrated to be effective in Japan as a way to avoid primary enucleation in Group D (Vb) eyes. This approach was modified slightly and popularized by Abramson and colleagues in New York. Later this morning we will hear the Italian experience with primary regional chemotherapy.

**INTRAVITREAL CHEMOTHERAPY (IViC):** The presence of diffuse vitreous seeding seems to be the component of intraocular disease that escapes control most commonly. This fact has led to the increasing use of direct intravitreous injection of melphalan (IViC) as described by Francis Munier, MD who is also presenting later this morning.

**VITRECTOMY WITH MELPHALAN INFUSION:** Recurrent and non-responsive vitreous seeding is also being treated with vitrectomy concurrently with infusion of melphalan in a large cohort of patients from all over China by Zhao Junyang, MD and his team in Beijing. Of their first 12 patients treated, 4 eyes were salvaged, two with good central acuity. Two children died of metastatic disease but there was no evidence of local escape of the tumor from the operated eye.

**PERIOCULAR INJECTION OF CARBOPLATIN/TOPOTECAN WITHOUT BARRIER CONTROL:** Injection of carboplatin into the subtenon space has been tried as primary therapy and to supplement CEV systemic chemotherapy in COG ARET0231 designed for Group C and D eyes. The COG study has been discontinued because of unacceptable side effects. Small patient series from Argentina and the experience in Toronto with periocular topotecan have not duplicated the complications seen with carboplatin. Sustained-release topotecan doses of up to 4 mg (in fibrin sealant) seem to be safe in animal models and in selected individual patients.

**DEFINING THE BARRIER TO TRANSSCLERAL DELIVERY OF THERAPEUTIC AGENTS:** Transscleral delivery of anticancer drugs, while promising theoretically, has been tried without great success. Iontophoresis did not improve transscleral drug delivery. In 2006, the barrier to transscleral drug delivery was shown to be dynamic, not static. Injected agents are rapidly washed out into systemic circulation by vessels and lymphatics in the conjunctiva. Efficient transscleral drug delivery requires that the concentration gradient across the eye wall be maintained.
**SEQUESTERED SUSTAINED-RELEASE TRANSSSLERAL DRUG DELIVERY:** In Southern California, we have worked in partnership with 3T Ophthalmics, Inc. to overcome the dynamic barrier by bringing to market a small silicone reservoir prefilled with a solid, sustained-release drug-matrix formulation of topotecan. When inverted and securely attached to the episclera with a bioadhesive, the reservoir sequesters the drug from access to lymphatics and conjunctival vessels so that drug washout does not occur. Water from inside the eye diffuses through the sclera and continuously and slowly dissolves the formulation releasing active topotecan to diffuse to the interior of the eye. Because of the sustained high concentration gradient, net diffusion of the drug is from the reservoir to the interior of the eye. The Episceral Reservoir delivers between 30 and 40 times more drug to the retina and vitreous than a does a similar amount of the drug injected into the subtenon space. FDA approval is expected within 45 days and Phase I studies will begin in Los Angeles.
L9 Imaging of paediatric ophthalmic tumors: towards new guidelines for MR imaging
Pim de Graaf
VU University Medical Center, Department of Radiology, Amsterdam, the Netherlands

This presentation will provide an overview of the diagnostic imaging modalities available and diagnostic strategy for the most common intraocular tumors and their main differential diagnoses. The most appropriate techniques for imaging in a child with leukocoria are reviewed. The historical utility of CT scan and its replacement by MRI as its imaging technology has overcome the early drawbacks of lack of spatial resolution and low signal-to-noise ratio will be discussed. The diagnosis of an intraocular tumor in a child is usually established by the ophthalmologist on the basis of fundoscopy and ultrasound. Together with ultrasound, high-resolution MRI has emerged as an important imaging modality for pretreatment assessment, i.e. for diagnostic confirmation, detection of local tumor extent, detection of associated findings. Minimum requirements for pretreatment diagnostic evaluation of intraocular tumors and mimicking lesions are presented. The potential value of advanced MR imaging techniques (Diffusion-, Perfusion- and Susceptibility-weighted imaging) for cancer imaging will be reviewed.
Management of intraocular retinoblastoma not only affects visual outcomes but also ocular retention and morbidity. Treatments include systemic and local chemotherapy, laser-, cryotherapy, external beam or plaque radiotherapy and enucleation. When treated, overall survival rounds 96% in well developed countries. In order to minimize the adverse events of these therapies and avoid enucleation for advanced retinoblastoma, a recent popularized technique is the use of chemotherapy agents delivered directly into the ophthalmic artery. Intra-arterial chemotherapy (IAC) is performed by interventional-neuroradiologists and is technically difficult. The treatment consists of administrating high doses of chemotherapy directly to the tumor, minimizing systemic absorption and drug-related systemic toxicity. The reduced systemic toxicity allows for the use of highly effective drugs against retinoblastoma, namely melfalan. Impressive tumor response of primary retinoblastoma with a high rate of global salvage has been reported with this technique. However, the success rate with IAC is lower for advanced disease where subretinal and / or vitreous seed recurrence can be a problem. There remain concerns about issues related to chemotherapy, infusion techniques and metastasis disease. IAC has an established role as salvage therapy and has been gaining ground as primary treatment for advanced retinoblastoma. We review and discuss current data regarding IAC for retinoblastoma and share our experience.
L12  Intra-vitreal treatment of retinoblastoma
Francis Munier
Jules-Gonin Eye Hospital, Lausanne, Switzerland
Mutations of both alleles of the RB1 gene that result in a loss of at least some of the diverse normal functions of the pRb protein stand at the beginning of the development of most retinoblastomas (Rbs). However, many Rbs show additional chromosomal aberrations and this suggests that Rb development is a multistep process. This view is also supported by genetic findings in retinoma, a non-proliferative retinal tumor. This possibly represents an intermediated stage of development and shows only few genomic changes other than RB1 gene mutations. Recurrent genomic changes typical for Rbs include gains of 1q, 2p and 6p, and loss of 16q and genes affected by copy number changes in these the regions have been proposed to be relevant for the progression of retinoblastoma. One of these genes, MYCN on 2p, shows moderate levels of gains in many Rbs with bi-allelic RB1 gene inactivation. The role of increased MYCN function for retinoblastoma development is highlighted by finding that some of the very rare Rbs that have normal RB1 (RB1+/+) genes show high level amplification of MYCN (MYCNA). Patients with RB1+/+/MYCNA tumors have isolated unilateral Rb and diagnosed at very young age. With new sequencing technologies (next generation sequencing, NGS) analysis of tumor genomes at the single base-pair level has become possible. In many tumors that occur in adults numerous genes are mutated and patterns of recurrently mutated genes have been identified. By contrast, so far NGS of Rbs has shown only few mutant genes other than the RB1. It has been proposed that, in Rbs, alterations of epigenetic regulation are more important for tumor development than copy number or sequence alterations. In fact, a comprehensive cross-data-type analysis on histone signatures, promoter methylations patterns, and gene expression revealed that activation of SYK gene expression has a prominent role for Rb progression. It is plausible that the epigenetic state of the Rb genome is a prerequisite for the malignant phenotype. Nevertheless, with only a few Rbs sequenced genome-wide, it is too early to dismiss the role of single gene mutations other that the RB1 gene.
Benign masses in childhood are generally either the result of developmental anomalies that may be evident at birth or may appear some time afterwards. Other childhood tumours are usually an abnormal proliferation of tissues in the growing child. The timing and treatment of benign disease in children is complicated by the risk of visual deprivation amblyopia, both before and immediately after therapy. Some benign tumours presenting in childhood, such as pleomorphic adenoma of the lacrimal gland, carry a risk of malignant transformation and dictate meticulous diagnosis and careful management. Examples of various benign masses presenting in the periocular region during childhood will be presented and the management of these conditions outlined.
L15  Towards reliable VEGF concentration measurements in blood samples of infants and young children
Birgit Lorenz, Claudia Lopez, Knut Stieger
Justus-Liebig-University Giessen, Dept. of Ophthalmology, Giessen, Germany

Speaker: Birgit Lorenz

Increased levels of vascular endothelial growth factor (VEGF) are thought to play a role in a number of pediatric retinal diseases, in particular primitive vascular abnormalities such as tumors and telangiectasias. Anti-VEGF treatment has therefore been used in various disorders, mostly anecdotically, and without solid data. The best studied entity today is retinopathy of prematurity (ROP) where increased intraocular concentrations of VEGF are a key factor in its pathogenesis. Measuring VEGF concentrations in different body fluids of preterm infants is therefore not only a major diagnostic parameter, but may provide crucial information during anti-VEGF treatment. In particular, the potential leakage of anti-VEGF molecules into the circulation and subsequent systemic down-regulation of VEGF, which might effect vessel maturation in other organs, needs to be monitored, making a robust and reliable measuring method clinically highly relevant. This talk will summarize the current state of the art in VEGF measurement and discuss possible ways to overcome existing limitations.
Support: Novartis Pharma GmbH, Germany
Different implant types and surgical techniques are available for enucleation in the paediatric patient. It is however not well defined which implant type or size, which surgical technique, or additional medical treatment is associated with a favourable outcome with respect to complications and cosmetic results. Furthermore, little is known about the quantity and type of functional problems associated with ocular prosthetic wear and/or external beam radiation in children. With our study group we are currently evaluating all patients that have been enucleated for retinoblastoma at the VU medical center since 1991. Some preliminary results are as follows: implant exposure, extrusion, socket abnormalities and infections of 225 enucleations were retrospectively analysed for association with implant type and additional therapy. We found that the combination of no implant and external radiation resulted in a contracted socket in 5 out of 8 cases (62.5%). Furthermore, the Allen-type implant tilted in 8 patients (8/18, 44.4%) and 5 needed to be replaced with another type of implant. The hydroxyapatite (HA) implants showed implant exposure in 8/82 (9.8%). Acrylic implants were exposed in 3/107 (2.8%). Implant extrusion was seen in 3/82 HA implants (3.7%), and 2/107 (1.9%) acrylic implants. Additional radio,- and/or chemotherapy in the HA/acrylic group (n=63) showed exposure in 7 (11.1%) and extrusion in 3 (4.8%) cases, while sockets without additional therapy (n=120) had a 3.3% exposure and 1.7% extrusion rate. Purulent discharge was noted in at least 39% of all sockets. It is concluded that HA and acrylic implant exposure and extrusion rates are rather small in our retinoblastoma population, with slightly higher exposure rates in the HA group in comparison to the acrylic group. Additional radio,- and/or chemotherapy results in a slight higher implant exposure and extrusion rate.
L17  Rhabdomyosarcoma  
Nathalie Cassoux  
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Orbital rhabdomyosarcoma (ORMS) is associated with an excellent survival rate greater than 85%, and considered as a favorable site for this tumor. The treatment is based on a poly-chemotherapy associated to the best local therapy, sometime surgery but more often radiation therapy, regarding to the initial tumor features and response. Local therapy is associated with frequent and potential severe late sequelae, thus pediatric oncology groups have tried over the years to reduce it without jeopardizing the outcome of ORMS with an adapted therapeutic strategy. A retrospective monocentric analyze on 95 patients will be shown.

These data suggest that orbital late effects are frequent after the treatment of an ORMS and lead to propose a systematic long term ophthalmologic follow-up of these patients. Radiation therapy is an important part of the total burden of therapy. Patients with a favorable pattern orbital RMS can be treated without radiation therapy in first line treatment in order to try to reduce local sequelae due to irradiation.
FP1 The ophthalmological examination in neurofibromatosis type 1

Sofie Caen\textsuperscript{1}, Ingele Casteels\textsuperscript{1}, Eric Legius\textsuperscript{1}, Catherine Cassiman\textsuperscript{1}

\textsuperscript{1}KU Leuven, Belgium

Speaker: Sofie Caen

\textit{Introduction}: The management, screening and follow-up of patients with neurofibromatosis type 1 (NF1) is still a debated topic. Several efforts have been made to standardize the ophthalmological examination. In 1997, the National Institutes of Health, NF1 Optic Pathway Glioma (OPG) Task Force, published a series of guidelines concerning the screening, monitoring and treatment of OPGs in NF1 patients. Listernick et al, members of the original Task Force, recently published some controversies and recommendations to their original publication. Little is known about the natural evolution of these low grade tumors and the effect of its growth or shrinkage on the ophthalmological symptoms. Therefore it is delicate to interpret the clinical findings and translate them into therapeutic decisions.

\textit{Objective}: The aim of this study is to compare several strategies of screening and follow-up in different ophthalmological institutes in Europe and United States of reference for NF1. We compare these results with the established screening guidelines as published by the OPG Task Force.

\textit{Methods}: A review of literature published between 1984 and 2013 was performed in search of screening strategies described in the articles. Methods of screening and follow-up were achieved through correspondence with different centers throughout Europe and the USA. After approval by the ethical committee a questionnaire was sent to these reference centers. This questionnaire consisted of ten questions about the ophthalmological, radiological follow up in their NF1 patients and of the genetic results if available.

\textit{Results}: We conclude that we are far from a consensus concerning ophthalmological screening and follow-up in NF1 patients. We will discuss these results to a greater extent and give some suggestions for future follow up schemes and its content. Only this will make it possible to achieve a better understanding of the natural history of OPG in NF1 and selecting patients that would benefit from therapy.
Optic Pathway Gliomas: Which role plays the ophthalmologist?
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Speaker: Yvonne Koenraads

Aim: To evaluate the role of the ophthalmologist in the process of early detection, diagnosis and follow-up of optic pathway gliomas (OPGs).

Methods: Patient files from all children with an OPG that were seen and registered in the University Medical Center Utrecht from 1994 up to 2012 were retrospectively reviewed.

Results: The patient population consisted of 25 children (15 ♂ + 10 ♀). OPGs were diagnosed at a mean age of 58 months (±SD 31 months) and included gliomas of the optic nerve (n=15), the chiasma (n=16) and/or the optic tract (n=5). Neurofibromatosis type 1 (NF1) was present in 17 children. In all children, diagnosis was based on MRI-imaging, subsequently, 6 children underwent an additional diagnostic biopsy. OPGs were diagnosed by a (pediatric) neurologist (n=13), an ophthalmologist (n=8) or a pediatrician (n=4). Moreover, the ophthalmologist had an essential contribution in the diagnosis in up to 52% of children. At moment of diagnosis, 18 children showed symptoms such as visual loss (n=8), headache (n=6), exophthalmos (n=2), epilepsy (n=1), ataxia (n=2), hemidystonia (n=1), reduced physical capacity (n=1), pubertas praecox (n=1), failure to thrive (n=1) and behavioral changes (n=1). Mean ophthalmological follow-up was 76 months (±SD 54 months). Visual outcome at the end of follow-up was normal in 7 children, while 18 children suffered from visual field (n=13) and/or visual acuity (n=12) deficits. Visual field assessment was not or could not be performed in 2 children.

Conclusion: The role of the ophthalmologist is of paramount importance in early detection, diagnosis and follow-up of OPGs. Standardized guidelines for neuro-ophthalmological follow-up of children with NF1 are needed for consequent screening on OPGs.
FP3  Tele-ROP for middle-income countries: 5 years of 'KIDROP' in India and the roadmap for universal infant eye screening
Anand Vinekar
1 Narayana Nethralaya Postgraduate Institute of Ophthalmology, Bangalore, India

Speaker: Anand Vinekar

Introduction: Over 3.5 million infants are born premature in India annually. With less than 20 ROP specialists nationwide, the vast majority of rural prematures are never screened and risk blindness. KIDROP (Karnataka Internet Assisted Diagnosis of ROP) initiated in Bangalore, India in 2008, addresses this problem. This presentation will highlight the lessons learnt in ROP screening and proposes the roadmap for universal screening from the data available.

Method: KIDROP employs a zonal approach of Tele-ROP screening using a portable wide-field digital pediatric retinal camera (Retcam Shuttle, Clarity MSI, USA), an indigenously developed hardware-software platform, trained technicians who screen infants in rural centres, on-site grading, live image reading and image analysis by the remote expert and real-time reporting using smartphones and tablet devices on a digitally secure platform within defined geographical areas where such facilities do not exist. The cost utility of this model in comparison with alternate methods was performed using analytic hierarchy processing (AHP).

Results: KIDROP currently provides ROP services to 81 neonatal units from 18 districts of Karnataka state in Southern India. Multiple teams have thus far imaged over 33,000 imaging sessions. Over 815 infants have undergone ROP treatment. Besides ROP, 7.2% of prematures were detected with other abnormalities. This led to a pilot of universal eye screening wherein 4.4% of healthy infants were serendipitously detected to have abnormal eye findings. The KIDROP model was ranked the highest on the AHP model compared to alternate methods.

Conclusion: KIDROP's model of remote screening using a tele-ophthalmology model has lessons for other middle-income nations, where the case load is high and trained specialists are few. Universal screening is promising, but the logistics are challenging and needs further validation.
FP4 The first results of the implementation of screening and treatment ROP in Astana

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¹ National Research Center of Maternity and Child Health, Astana, Kazakhstan

Speaker: Gulnara Utegenova

Introduction: Since 2011 In «NRCMCH» has started the scientific project which included the introduction of screening and treatment of threshold ROP stages. Ophthalmologists from Center organized the inspections of all preterm infants and attendances in offices newly born care clinics of Astana.

Results: As a consequence in 2011 the percentage of detection of active ROP stages in NRSMCH" achieved 17% in comparison with 2,3% in 2010, whereas in 2012 it reached 34,6%. In 2012, we have screened in 656 infants. The active stage of ROP was diagnosed in 228 children. Since the beginning of 2012 the ophthalmologists of center began to perform retinal laser coagulation independently. In case of progression of the disease to type 1 ROP, the transpupillary laser coagulation of avascular retina was performed for 83 children. We performed laser treatment on 164 eyes under endotracheal anesthesia. Due to the ongoing pathology in 9 infants we have repeated laser coagulation of the retina on 11 eyes.

Conclusion: In 2012 the efficiency of laser treatment was 94.7%. The introduction of international experience of timely treatment prevents the end-stage of ROP which leading to blindness."
FP5  Visual functions at 2.5 years in two cohorts of prematurely-born children in Sweden

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³ Department of Ophthalmology, Linköping University, Sweden,
⁴ Department of Ophthalmology, Lund University Hospital, Sweden

Speaker: Gerd Holmström

Introduction: The aim of the study was to investigate the visual and ophthalmic outcome in prematurely born children in Sweden at 30 months corrected age. Methods: Two Swedish population-based, prospective studies were initiated in 2004. One cohort included all extremely preterm (< 27 w GA) infants born in Sweden 1 April 2004 to 31 March 2007 (the EXPRESS study) and another comprised 111 very preterm children (<32 w) born 1 Jan 2004 -31 Dec 2007 in Uppsala county (the LOVIS study). Ophthalmic and orthoptic evaluations were undertaken at 30 months corrected age (CA).

Results: Eighty-four percent (411/491) of the surviving children of the EXPRESS study and 88% (98/111) of the LOVIS study took part in the follow-up at 30 months CA (+/- 2 months). The incidence of ROP was 73% in the national cohort (< 27w) and 25% in the Uppsala cohort (<32w). Treatment for ROP, according to ETROP criteria, was performed in 20% versus 6% in the two study groups. At 30 months CA, four children in the EXPRESS study, but none in the LOVIS study, were blind. Strabismus was found in 14% and 5% and refractive errors, defined as myopia < - 3D, hypermetropia > + 3 D, astigmatism ≥ 2 D, and/or anisometropia ≥ 2D, in 26% and 11% in the two studies, respectively.

Conclusion: As expected, visual and ophthalmic dysfunctions were most pronounced in the most prematurely born children. Whether the findings at 30 months corrected age will predict problems at school age will be investigated in on-going studies of both populations at 6.5 years of age. At that age a control group of children born at term are also investigated, making it possible to compare the results with those of the prematurely born children.
FP6  Pediatric corneal collagen cross-linking (CXL)
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Speaker: David Tabibian

Introduction: To study the progression rate of keratoconus and to assess the clinical outcome of corneal collagen cross-linking with riboflavin and UVA (CXL) in children and adolescent patients.

Methods: Fifty-nine eyes of 42 children and adolescents (aged 9 to 19 years) with confirmed keratoconus were included in this study. Fifty-two eyes showed progression and 46 eyes were treated with CXL. Refraction, slitlamp examination, placido-based corneal topography and Scheimpflug imaging were performed bilaterally in all patients preoperatively and at six and twelve months postoperatively. Kmax (maximal K readings), CDVA (corrected distance visual acuity), corneal thickness and the keratoconus index (KI) were analyzed. Follow-up was up to 3 years.

Results: Fifty-two of the 59 eyes enrolled in this study showed progression, corresponding to a progression rate of 88%. Forty-six eyes were treated by CXL. All parameters investigated showed significant improvements over the follow-up period. However, the significant Kmax reduction observed up to 24 months after CXL showed a trend towards regression at 36 months.

Conclusion: CXL seems to be safe in children and adolescents. Natural progression of keratoconus occurred in 88% of children and adolescents. We propose that awaiting documentation of progression is not mandatory and that CXL in children and adolescents should be performed as soon as the diagnosis has been made. However, the effect of arrest of disease progression might not be as long-lasting as in adults and longer follow-ups are needed to verify this trend.
FP7  Next generation sequencing as a powerful means of diagnosis for Congenital Cataract
Rachel Gillespie¹, Simon Ramsden², Ian Christopher Lloyd³, Graeme Black¹
¹ Centre for Genomic Medicine, Institute of Human Development, Faculty of Medical and Human Sciences, University of Manchester, MAHSC, Manchester, UK, ² Centre for Genomic Medicine, Central Manchester University Hospitals NHS Foundation Trust, Manchester Academic Health Sciences Centre (MAHSC), Manchester, UK, ³ Manchester Academic Health Science Centre, Manchester Royal Eye Hospital, Central Manchester Foundation Trust, The University of Manchester, Oxford Road, Manchester, UK

Speaker: Rachel Gillespie

Introduction: Congenital cataract (CC) is a major cause of blindness in children worldwide. Genetic mutations account for 50% of cases. More than 110 genes have been identified as causal. Early diagnosis is critical for clinical management but current investigations to determine aetiology are expensive and delay accurate diagnosis. Determining the precise genetic cause clarifies inheritance pattern and serves as an effective means of disease delineation in patients with syndromic CC. Genetic heterogeneity and technological limitations have reduced ability to diagnose and treat patients effectively, and have restricted understanding of the epidemiology of CC. Next generation sequencing (NGS) technologies are likely to address these issues.

Methods: We have designed an NGS target enrichment that permits the analysis, in parallel, of 113 genes known to be mutated in CC.

Results: Results show NGS technologies are able to accurately and efficiently determine the precise genetic cause of CC in 70% of individuals. More than 80% patients with isolated CC were found to have apparently pathogenic mutations, all of which occurred in highly conserved domains known to be vital for normal protein function. The pick-up rate in patients with syndromic CC was also high, with 56% found to have potential disease causing mutations. In this small cohort, the array has identified some rare causes of CC.

Conclusion: These results demonstrated the significant clinical utility of this test including examples altering clinical management, directing care pathways and enabling more relevant counseling. Establishing more robust genotype-phenotype correlations and advancing our knowledge of cataract forming mechanisms will aid research into targeted therapies. We believe that this comprehensive screen will extend access to genetic testing and will lead to improved diagnostic outcomes attributable to the implementation of stratified medicine.
FP8 The role of autoimmune mechanisms in the development of paediatric chronic recurrent uveitis

Ganna Lysenko

1 Donetsk National Medical University, Department of Ophthalmology of Faculty Postgraduate Education, Donetsk, Ukraine

Speaker: Ganna Lysenko

Introduction: Uveitis in children is a very severe disease, more likely to lead to vision loss and in most cases characterized by chronic recurrent course as well as high frequency of complications. The purpose of the study was to investigate the role of autoimmune component in the development of chronic recurrent uveitis in children, by determining systemic autoantibodies as well as specific autoantibodies against structural components of eyes.

Methods: Serum of 15 patients with chronic recurrent uveitis aged from 6 to 18 were investigated by method of indirect immunofluorescence on the presence of the systemic autoantibodies against cell nuclei (ANA) and organ-specific autoantibodies against structural components of eyes on sections of monkey eyes. The patients also were examined by IFA on presence of the autoantibodies against cytoplasmic proteins of neutrophil granulocytes-myeloperoxidase (MPO) and proteinase 3 (PR3).

Results: All patients have chronic recurrent course of disease with duration from 1 month to 5 years and the number of recurrences is from 2 to 5 per year. Bilateral cases were revealed in 36% children. The leading form of uveitis in the study group were posterior uveitis in 8 patients (60%), while anterior uveitis were observed in 5 patients (36%). ANA were detected in 71% patients, including high level in 36%, average level in 21%, threshold in 14%, not diagnostically relevant in 7%. Autoantibodies against cytoplasmic proteins of neutrophil granulocytes were detected in 14% cases. In the vast majority of patients with chronic recurrent uveitis were observed specific autoantibodies against structural components of eyes, including autoantibodies against outer and inner nuclear layer of retina in 92,6 %, against rods in 71,4%, against tunica choroidea in 78,6%, furthermore, against endothelium of vessels in 14% cases and against myelin in 35,7% patients. No case detected autoantibodies against cones. It should be noted that some cases have positive results of specific autoantibodies against structural components of eyes and meanwhile have negative results of systemic ANA.

Conclusion: Autoimmune mechanisms play crucial role in pathogenesis of chronic recurrent uveitis in children. Examination of the children with chronic recurrent uveitis should include the investigation of systemic autoantibodies as well as organ-specific autoantibodies against structural components of eyes that determines further treatment of such patients.
FP9  Artificial Iris Implants for Aniridia and Oculocutaneous Albinism: Near-Infrared Light Transmission May Risk Retinal Phototoxicity and Cataractogenesis
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Speaker: Imran Yusuf

Introduction: Artificial iris implants may be implanted in patients with congenital aniridia and oculocutaneous albinism to restore iridocosmesis and the light-occlusive properties of the iris pigment epithelium. Spectral transmission analysis has identified high levels of near-infrared (NIR) light transmission through some black intraocular lenses. Artificial iris implants aim to produce light occlusion from similar materials but spectral transmission is unknown. Incomplete light occlusion may risk long-term adverse biological effects on ocular tissues in this patient group, preventable by implantation of NIR-blocking iris implants.

Methods: Spectral transmission profiles of Morcher Black Diaphragm implants (BDI; 67B, 68, 67G, 67F, 67L, 94A) and Dr Schmidt’s Artificial Iris implants (hazel and brown iris illustrations) were acquired using a broad-spectrum light source and spectroradiometer. Transmission data was used to model the NIR light transmitted as a percentage of total retinal incident light (photons) under photopic conditions, based on daylight, fluorescent and incandescent light sources.

Results: Spectral transmission revealed total occlusion of light by Dr Schmidt’s Artificial Iris implants. All Morcher BDI iris implants demonstrated high levels of NIR transmission across the black iris interface. NIR light transmitted across the Morcher BDI interface as a percentage of total retinal incident light was dependent on the diameter of the BDI pupillary aperture: Morcher 67B BDI (3mm pupil diameter) transmitted 34% of fluorescent light, 61% of photopic daylight, and 82% of incandescent light through the NIR window, versus 6%, 17% and 37%, respectively, for the Morcher 94A BDI implant (6.5mm pupil diameter).

Conclusions: Dr Schmidt’s Artificial iris implants produce physiological light occlusion identical to that of the native human iris. However, Morcher BDI iris implants transmit nearly 100% of near infra-red light, exposing the posterior segment to chronic excess NIR light. Artificial iris implants are intended for long-term iris restoration, increasing the cumulative preventable NIR light exposure and risk of adverse biological effects on ocular tissues. NIR light is known to produce retinal injury and accelerate cataract formation. NIR-blocking Artificial iris implants may represent a safer and superior physiological iris prosthesis compared to NIR transmitting iris implants in this patient group.
FP10 The uptake of prenatal testing for retinoblastoma susceptibility by pregnant women at increased risk

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Speaker: Charlotte Dommering

Introduction: Prenatal diagnosis (PND) is discussed with all couples at risk of having offspring with hereditary retinoblastoma, based on a previously identified germline mutation of the RB1-gene. Couples have the option to terminate the pregnancy, if the foetus tests positive. The objective of this study was to evaluate the nationwide uptake and outcome of PND for this cancer susceptibility.

Methods: We scored the total number of families with a known germline mutation of the RB1-gene and of requests for PND. Follow-up data were obtained by reviewing medical files. Uptake of PND for retinoblastoma susceptibility was compared with other hereditary high-risk cancer susceptibility syndromes, nationwide.

Results: We identified 186 families with a known RB1-mutation in the Netherlands. PND was performed 36 times for 23 families (12%). Fifteen couples had a recurrence risk of 50% and eight a recurrence risk of 2-3% (based on possible germline mosaicism). Eleven foetuses tested positive; 8 pregnancies were terminated. Of the 365 nationwide registered families with familial adenomatous polyposis with a germline mutation of the APC-gene, 9 (2%) performed PND. Likewise, PND was performed in less than 0.2% of > 3000 nationwide registered BRCA1/BRCA2-mutation positive families and in five of 48 registered families with Von Hippel Lindau syndrome (5%).

Conclusion: Hereditary retinoblastoma has a high impact on reproductive decisions. Compared to other hereditary cancer syndromes, prenatal diagnosis seems to be performed substantially more often for retinoblastoma susceptibility. Possible reasons for differences will be discussed.
FP11 Retinal detachment after retinoblastoma treatment
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Speaker: Pascal Dureau

Introduction: Conservative treatments for retinoblastoma allows anatomical and sometimes functional rescue of the eyes. However, some of these eyes experience early or late retinal complications: radiation retinopathy with neovascularization, vitreous hemorrhage, retinal atrophy, that can lead to tractional or rhegmatogenous retinal detachment. The aim of this study was to evaluate the clinical characteristics, treatment and results of retinal detachment in eyes previously treated for retinoblastoma.

Methods: Retrospective study of children operated for retinal detachment after retinoblastoma treatment from 1998 to 2011. The treatment used for the tumor, the time before onset of detachment, the type of detachment, the type of surgery, the anatomical and functional results were noted.

Results: Ten children were operated during this period. All had bilateral retinoblastomas with the fellow eye enucleated. Five had been treated by a combination of radio and chemotherapy, three by thermochemotherapy and two by radiotherapy. Seven detachments were tractional, and three rhegmatogenous.

Median time after initial treatment was 52 months (3-200). Seven eyes were treated by scleral buckling, nine by vitrectomy, three by silicone and three by gas tamponade. The retina was reattached in six cases. Mean follow up was 26 months. Final visual acuity was 20/40 in 2 cases, 20/200 in one, hand movements in 3 and no LP in 4. No child experienced recurrence of the tumor.

Conclusion: The mechanism or retinal detachment after retinoblastoma treatment can be tractional, generally associated with radiation retinopathy, neovascularization and vitreous hemorrhage, occurring relatively early after treatment, or rhegmatogenous, related to atrophic holes at the edge of the residual tumor. The decision for treatment must be carefully considered, regarding the risk of recurrence or dissemination and the difficulty of surgery. Eligible cases are mostly bilateral after enucleation of the fellow eye, and the visual prognosis remains limited.
FP12 Evaluating the risk of extraocular tumor spread following intravitreal injection therapy for retinoblastoma: a systematic review

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Speaker: Stephen Smith

Introduction: Intravitreal injection therapy (IViT) for retinoblastoma has shown promise in the treatment of vitreous seeds; however, the potential for tumor dissemination following intravitreal penetration has limited its use. This review evaluates the risk of extraocular tumor spread in patients receiving therapeutic intravitreal injections for retinoblastoma.

Methods: PUBMED (1946-present), SCOPUS (all years), Science Citation Index (1900 – present) and Conference Proceedings Citation Index – Science (1990 – present) electronic databases were searched to identify all published reports of IViT for retinoblastoma in humans.

Results: Fourteen studies with original IViT data were included in this review. A total of 1304 intravitreal injections were given in 315 eyes of 304 patients, with one report of extraocular tumor spread and one patient in which intravitreal treatment could not be excluded as a contributor to metastatic disease. The proportion of subjects with extraocular tumor spread potentially due to IViT in these combined reports was 0.007 (95% confidence interval (CI): 0.0008 to 0.0236), with a mean follow-up of 72.1 months. In a subset of 61 patients receiving IViT via safety enhancing injection techniques (347 injections, 19.6 mo mean follow-up), there were no reports of tumor spread.

Conclusion: Local and systemic tumor spread following intravitreal injection therapy in cases of retinoblastoma is rare, and this risk is potentially reduced by the use of safety enhancing injection techniques. These results suggest that the risk of tumor spread should not preclude IViT use for carefully selected patients as part of multimodal globe salvaging therapy.
Purpose: To evaluate visual outcomes, tumor recurrences and the clarity of the visual media to check the ocular fundus among children with retinoblastoma undergoing limbal cataract aspiration and intraocular lens implantation for cataract secondary to external beam radiotherapy.

Methods: This is a retrospective review of the medical records for all patients who underwent limbal cataract aspiration and in-the-bag IOL implantation for cataract secondary to external beam radiotherapy for retinoblastoma, between January 2001 to May 2013. Main outcome measures were: Interval from last retinoblastoma treatment and cataract surgery, pre and postoperative visual acuity, clarity of visual axis to check ocular fundus and tumor recurrence.

Results: The study included 13 patients, 10 with bilateral and 3 with unilateral retinoblastoma. Of these 23 eyes, 16 eyes had cataract surgery, 7 eyes were enucleated, and 1 eye required enucleation after the surgery for tumor recurrence. The mean follow-up after surgery was 58 months (range 3 months-146 months). The quiescent interval before cataract surgery was between 12 and 36 months, except one patient who had surgery 12 years after last retinoblastoma treatment. A laser capsulotomy was subsequently performed in 5 eyes. Final visual acuity was between 0,6 and 1 in 6 eyes, between 0,2 and 0,4 in 2 eyes and equal or worse than 0,05 in 7 eyes, with 8 eyes having macular tumor involvement. In all patients, the media were clear enough to check the fundus after cataract surgery. We present Ret-Cam pictures of the fundus, before and after cataract surgery.

Conclusions: Limbal cataract surgery without posterior capsulotomy is a safe procedure for radiation-induced cataract and allows visual acuity improvement and clear media to check the tumors.
FP14 Direct intraarterial (ophthalmic artery) chemotherapy for advanced intraocular retinoblastoma: 5 years' experience
Theodora Hadjistilianou¹, Sonia & Maria Carla De Francesco & De Luca¹, Paolo Nucci³, Sandra Bracco², Paolo Galuzzi², Carlo Venturi³
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Speaker: Doris Hadjistilianou

Objective: Report five years of conservative treatment for advanced retinoblastoma with direct intraarterial-ophthalmic artery infusion of Melphalan alone or Melphalan and Topotecan.

Materials: 75 children (82 eyes) with advanced retinoblastoma (Stage D-E/VA –VB) in phase two of one center. Seven treated bilaterally. 40 eyes were first diagnosis (naïve) and 42 were relapses following chemotherapy and focal therapy and/or radiotherapy.

Methods: Cannulation of the ophthalmic artery was performed by a femoral artery approach using microcatheters (magic 1.5) under general anesthesia and anticoagulated. Chemotherapy (Melphalan alone or Melphalan and Topotecan) was infused into the artery over a 30-minute period (dose of 3-7 mg of Melphalan and 0.3-0.4 mg of Topotecan, according to the age and size of the globe). Local and systemic toxicity were documented.

Results: The ophthalmic artery was successfully cannulated in 82 eyes (total, 390 procedures). Complete remission was achieved in 26 out of 40 naïve cases. 14 eyes out of 40 were enucleated. In the group of the relapsed eyes 28 out of 42 achieved complete remission. 14 out of 42 underwent enucleation for insufficient of response. There were no strokes, seizures, dissections, carotid and femoral arteries occlusions or bleeding, central retinal arteries spasms or occlusions, cardiac arrests, and admissions in neurointensive care unit following the intra-arterial procedure. One patient underwent blood transfusion for the worsening of preexistent chemotherapy-related anaemia. Neutropenia (1 grade 1–2; 1 grade 2), occurred in the two patients with 13q deletion syndrome. Follow-up range is 1-60 months. Ocular adverse events included eyelid oedema (39 cases), ptosis (1 case), nasal loss of lashes (3 cases), frontal alopecia (2 cases) and frontal rash (10 cases), strabismus: exotropia (1 case). Vascular adverse effects included: acute ischemic papillitis, Roth spots and foveolar haemorrhage. Eight patients developed progressive chorioretinal atrophy associated with RPE damage. There was no toxicity to the anterior segment.

Conclusions: 75 children (82 eyes) with advanced retinoblastoma were eligible for the intra-arterial Italian Protocol. The 65.8% of all treated eyes is in complete remission. Superselective chemotherapy delivered through the ophthalmic artery can avoid enucleation, primary radiation or abuse of systemic chemotherapy.
FP15 Suspected optic disc oedema in children – how helpful is ultrasound ONSD measurement in clinical decision making?
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Speaker: Anne Cees Houtman

Introduction: Assessment of the paediatric optic nerve head in the context of possible raised intracranial pressure (ICP) can be difficult. Assessment of disc oedema is dependent on the cooperation of the child and observer variation. Furthermore, disc oedema may be absent in the presence of raised ICP. The consequences of a missed diagnosis of raised ICP may add to an ophthalmologist’s uncertainty and lead to unnecessary referrals and more diagnostic tests like neuroimaging and lumbar puncture. Ultrasound examination may be used to assess the presence of increased optic nerve sheath diameter (ONSD) which correlates well with raised ICP.

Methods: To assess the value of ultrasound-Optic Nerve Sheath Diameter (US-ONSD) measurement in cases of suspected paediatric optic disc oedema we reviewed a cohort of children in a large university medical center in the north of the Netherlands. A large US-ONSD database was manually searched for cases (age range 0-16 yrs) in which the aspect of the optic disc – assessed during routine clinics – raised suspicion of disc oedema and for whom an US-ONSD measurement was requested for confirmation of distension of the optic nerve sheath (ONS). Paediatric (non-ophthalmic) case notes were scrutinized for missed diagnoses that came to light after ONSD measurements.

Results: Out of several hundred ONSD measurements we identified 98 children with suspected disc oedema assessed by ophthalmologists, ophthalmology trainees or paediatric neurologists (e.g. crowded, tilted discs, papilloedema etc). Mean follow-up was 36 months (range 1-76). In 30 cases the quantitative assessment of the ONSD was higher than normal (>5.0 mm). In 26 of these other specialists, mostly paediatric neurologists, were involved in the management (4 were not referred, 3 were lost to follow-up). 10 of these patients had conditions with raised ICP. The qualitative assessment of the ONS (doughnut, hanging or flying bat) in these patients was very suggestive of ONS distension in 6 cases, moderately so in 3 and not suggestive in 1 (though it quantitatively was). In the remaining 68 cases with ONSD measurements in the normal range only 1 (with marginal ONSD of 4,95 and 5,0 mm) was found to have raised ICP. During follow-up no cases of a missed diagnosis of raised ICP came to our attention.

Conclusion: US-ONSD measurement appears a safe and useful tool to assess raised intracranial pressure in children with suspected optic disc oedema.
FP16 Histiocytic lesion of the orbit in children

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¹Hospital ant Joan de Deu de Barcelona

Speaker: Joan Prat

Introduction: The more frequent histiocytic orbital lesions in children are Langerhans cell histiocytosis (HCL) but don’t the only one. We compare clinical manifestations of histiocytic lesions depending on age.

Methods: Twelve orbital histiocitic lesions in children from 9 month to 10 years seen during the last 10 years in a tertiary department of pediatric ophthalmology. We have recorded the clinical manifestations, CT/MRI and therapy. The compared parameters are summarized in: diagnosis of the disease, extension and response to treatment.

Results: HCL was found in 9/12 cases. Orbital roof involvement appeared as a eyelid subacute edema associated to a osteolitic lesion on CT. Skull and vertebral bones are the most affected. Infrequent disorders were Erdhein Chester disease, a micobacterial infection and a hypereosinophilic inflammatory lesion.

Conclusion: Younger children with HCL and orbital involvement present more systemic extension than the older children. Others histiocytic lesions must be considered.
FP17 Morbus Graves in children and adolescents
Irene Notting¹, Nicoline Schalij-Delfos¹
¹ Leiden University Medical Center, the Netherlands

Speaker: Irene Notting

Introduction: M Graves of the eye’s is a rare disease in children and adolescents. Methods: We describe a retrospective study of 16 patients with M Graves which visited our clinic between July 2009 and July 2013.

Results: Fourteen girls and two boys visited our clinic with a mean age of 16.4 years (11-22 years). Two patients (13%) had no ophthalmic signs of graves orbitopathy (GO), 62% had Nospecs 1-3 (mean age 15.8 years (11-21)) and 25% had Nospecs 4-6 (mean age 19.5 years (15-22)). Radiologic exams (MRI/CT orbit) were performed in 7 patients, 5 showing pathology due to M Graves. One patient was treated with steroids (Amsterdam protocol) and one patient was decompressed surgically. Most patients (12) were treated with block and replacement therapy, two were operated and two got radioactive iodine.

Conclusion: GO developed in patients above 15 years. No muscle enlargement or lipid hypertrophy was found in children under 15 years. From the age of 15 regular check-up will be necessary because severe GO can develop.
FP18 Melanocytic tumors and their look alikes in and around the eye
Evelyne van de Winkel, Irene Notting, Marina Marinkovic, Gregorius Luyten, Nicoline Schalij-Delfos
1 Leids Universitair Medisch Centrum, the Netherlands

Speaker: Evelyne van de Winkel

Goal: Inventory of melanocytic neoplasms in children, followed or treated since 2008.
Method: Retrospective study of infants born after 1990 with a registered diagnosis tumor/neoplasms in the hospital registration system. Evaluation of history, age, clinical diagnosis, treatment, documentation and histopathological diagnosis.
Results: Inclusion of 109 children with melanocytic neoplasms (0-21 yrs): 57 conjunctival nevi, 8 iris nevi, 3 choroideal nevi; 6 ocular melanosis; 4 hamartomas; 1 primary acquired melanosis (PAM); 2 choroidal melanomas, 1 iris melanoma suspect and 27 others. Of the 68 nevi, 66 were photographically documented showing a large number of clinical variations. Excision of 35 conjunctival nevi because of growth (10), change in aspect (12), irritation (4), cosmetic (1) or not documented (8). Melanomas were excised or treated with Ruthenium plaque therapy and the other melanocytic lesions were followed closely. Histopathologic diagnosis of the nevi revealed 2 types: melanocytic andnevocellular nevus, the excised choroidal melanoma was of the mixed cell type and the iris melanoma biopsy showed atypical melanocytic proliferations and focal necrosis suspect for malignancy.
Conclusion: With exception of 3 malignant melanomas, most melanocytic tumors in this group were benign neoplasms, especially conjunctival nevi. Although the diversity in presentation of these nevi was large, the histopathological diagnosis in all cases was benign.
FP19 Clinical features, instrumental diagnostics and surgical treatment of orbital rhabdomyosarcoma in children
Svetlana Saakyan¹, Anush Amuryan¹, Valida Alichanova¹, Olga Ivanova¹
¹ Moscow Helmholtz Research Institute of Eye Diseases, Russian Federation

Speaker: Svetlana Saakyan

Introduction: To present the clinico-instrumental features and surgical approaches in orbital rhabdomyosarcoma in children.

Methods: 15 children in the age from 4 month to 14 years (4 girls and 11 boys) were treated in our clinic. All the patients underwent both clinical and instrumental examinations, including CT-scan, high frequency ultrasonography. The surgical treatment was performed in all cases – tumor excision or incision biopsy. The morphological verification of the tumors’s type was dedicated in all children: embryonal (13 children) and alveolar (2 children) rhabdomyosarcomas. Afterwards all children underwent both radiotherapy and chemotherapy.

Results: In all case the progressive proptoses with vision decreasing in short period of time were noticed (from 2 weeks to 4 month). CT scan demonstrated a solid mass with accurate margins, localized in different part of the orbit (mainly under the upper roof – in 6 cases, rare in medial (in 1 case) and lateral (in 1 case) parts) with infiltration of rectus or oblique extraocular muscles. High frequency ultrasonography with Doppler scanning showed a hyporeflective lesion with hypervascular internal vascularization in all the tumors. In 6 cases the tumor excision were performed (all these tumors were covered with thin pseudocapsula and has less infiltration of orbital tissues and extraocular muscule).

Conclusion: The examination of rhabdomyosarcoma should be complex, based on anamnesis data, clinical features and instrumental investigations results. The type of surgical treatment (tumor excision or incision biopsy) should be chosen depending on the tumor infiltration of orbital tissues and possibilities to remove the tumor mass without the breaking of tumor’s pseudocapsula.
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RAPID FIRE
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POSTER
RF01 Functional visual loss or brain tumour? The use of sweep VEP
Maria van Genderen¹, Ymkje Hettinga¹, Frans Riemslag¹, Gerard de Wit¹
¹Bartiméus Institute for the Visually Impaired, the Netherlands

Speaker: Maria van Genderen

Introduction: The estimation of visual acuity via visual evoked potentials (VEP) can be assessed with a spatial frequency sweep, utilising check sizes from 60’ to 1.5’. Sweep VEP is a valuable objective measure in patients suspected of functional visual loss.

Case reports: In a short period of time, we saw two nine year old girls who both complained of visual loss since several weeks. BCVA was 1/300 in girl 1 and 1/60 in girl 2. Peripheral visual fields were normal; colour vision could not be tested. Both children had normal pupillary responses, a normal fundus without optic disc abnormalities, and a normal OCT. Both girls were therefore suspected of functional visual loss. Sweep VEP in girl 1 showed recordable responses up to 1.5’ check size, indicating normal vision. However, in girl 2 no VEP responses could be measured. Because of this unexpected finding, we investigated whether visual behaviour, for instance not fixating centrally, could have influenced the results. The investigation showed that fixation just beside the screen resulted in reduced response amplitudes, but that responses still could be recorded. We therefore referred girl 2 for neurologic evaluation. On MRI, she proved to have a craniopharyngeoma. After surgery, visual acuity returned to normal, although she had a remaining visual field defect.

Conclusion: Sweep VEP can be used to discriminate between functional visual loss and visual loss from organic causes. As children with craniopharyngeoma may have normal pupillary reflexes and normal optic discs, abnormal VEP responses may be the only indication of a serious neuro-ophthalmologic disorder. On the other hand, a normal sweep VEP may reduce the amount of unnecessary and costly diagnostic procedures (MRI, LP, etc) in children with functional visual loss.

See also: Poster P02.
RF02 Efficacy and safety of a combination of 0.5% Cyclopentolate with 2.5% Phenylephrine used for ROP screening in 623 consecutive examinations spanning a 30 month period
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1 Royal Berkshire Hospital, Reading and King Edward VII Hospital, Windsor, UK

Speaker: James Neffendorf

Introduction: Neonatal screening for Retinopathy of Prematurity (ROP) requires efficient mydriasis with minimal unwanted side effects. The current Royal College of Ophthalmologists, UK, (RCOphth) guidelines advocate a mydriatic combination of 0.5% cyclopentolate with 2.5% phenylephrine, instilled 2-3 times, 5 minutes apart. However, no large studies have been performed to validate this combination in terms of safety and efficacy. Small case series have shown associations with apnoeic episodes and gastro-intestinal disturbance.

Methods: 623 consecutive examinations (1,246 eyes) for babies undergoing ROP screening by the same examiner (GDH) were analysed for mydriatic efficiency and severe complications during and within the first 24 hours after examination.

Results: 98.8% of cases achieved successful mydriasis to complete full examination. No severe intra-procedural problems requiring cessation of screening were encountered. Within the subsequent 24-hour-period, we found 5 cases with apneic episodes requiring medical intervention and one case of necrotizing enterocolitis 1 week post-screening. As there was no control group, it is unclear whether these cases represent an increased risk. None of these children demonstrated repeat apnoea or other significant systemic complications if redilated subsequently.

Conclusion: We believe this is the first report of a large cohort of the effectiveness and safety of combined 0.5% Cyclopentolate and 2.5% Phenylephrine for ROP screening in premature babies.

See also: Poster P07.
RF03 Long term refractive status follow-up of prematures: singletons vs multiples
Hana Leiba¹, Niv Levy², Eric Shinwell¹
¹ Kaplan medical center, Rehovot, Israel, ² Hadassah Medical School, the Hebrew University, Jerusalem, Israel

Speaker: Hana Leiba

Background: Premature birth is associated with a greater incidence of refractive disorders, both with and without Retinopathy of Prematurity (ROP). In addition, multiple gestations have been associated with increased ocular morbidity.

Aim: To compare the refractive status at 8-12 years in preterm and term-born children and in single and multiple pregnancies. To assess the effect of potential risk factors and refractive status at age 6 months on refractive status at 8-12 years.

Methods: Children from either single or multiple births, with birth weight less than 1500 grams, who had documented ophthalmic examination at six month corrected age, were studied. The control group included age matched children who were born at term.

Results: 37 of 104 preterm infants were from multiple pregnancies. At six months, most were emmetropes and hyperopes. At 8-12 years, although most children were emmetropes, multiples had significantly more myopia (S 13% Vs M 38%).

Multivariate regression analysis revealed a significant relationship between refraction at age six months and multiple birth status and refraction at 8-12 years. Birth weight and the presence of ROP were not statistically significant in this model.

Conclusions: Refraction at age 6 month is strongly correlated with refraction at age 8-12 years. Preterm children have more refractive errors than those born at term. Multiple gestation, independent of prematurity, increases risk for refractive errors, particularly myopia.

See also: Poster P08.
RF04 Macular function measured with mfERG in prematurely-born children at school-age  
Hanna Åkerblom¹, Gerd Holmström¹  
¹ Dep. of neuroscience, Uppsala University, Sweden
 Speaker: Hanna Åkerblom

Introduction: Children born prematurely have affected visual functions compared to children born at full-term, including decreased visual acuity and contrast vision. The aim of this study is to compare macular function in children born preterm with full-term children. A second aim is to correlate macular function with gestational age (GA), birth weight (BW), visual acuity (VA) and macular thickness.

Methods: Nine children, 9-13 years old, born before 32 gestational weeks, was evaluated with multifocal Electroretinogram (mfERG) and Optical Coherent Tomography (OCT). Mean GA was 29 weeks and mean BW was 1259 g. Four children had no ROP and five had mild ROP in the neonatal period. MfERG results, from the right eye of eight full-term children, 8-19 years old, with normal visual acuity, acted as control group.

Results: The amplitude of the P1 response of the mfERG was significantly reduced compared to controls. The implicit time of the P1 response showed no difference between the two groups. There was no correlation between P1 amplitude and GA, BW, VA or central macular thickness in the preterm group.

Conclusion: The macular function measured with mfERG is affected in prematurely-born children at school-age, with no or mild ROP in the neonatal period. These preliminary results show no correlation between macular function and visual acuity or central macular thickness. Previous studies have shown increased macular thickness in prematurely born children suggesting a disturbed development of the macular morphology. The results in the present study show signs of a disturbance in the macular function as well as the macular structure; this may be one of several reasons to reduced visual acuity in children born preterm.

See also: Poster P09.
RF05 Twins with bilateral posterior microphthalmos
Yvette Braaksma-Besselink, Marije Sminia, Linda Groenveld
1 Academic Medical Centre (AMC) Amsterdam, the Netherlands

Speaker: Yvette Braaksma-Besselink

Introduction: Posterior microphthalmos is a rare congenital bilateral eye condition in which the posterior segment is abnormally small. The most striking symptom is a high hypermetropia with typical retinal abnormalities and a short axial length.

Methods: Case report, we describe identical twins in whom suboptimal visual acuity with optimal refractive correction and originally normal fundus led to more extensive research and ultimately to the diagnosis posterior microphthalmos. The prognosis regarding visual acuity varies per patient. There is a risk of uveal effusion and/or acute closed angled glaucoma. Sometimes combined with retinopathy.

Conclusion: Be aware of posterior microphthalmos in patients with suboptimal visual acuity with high hypermetropia. Yearly checkup by an ophthalmologist is a necessity.

See also: Poster P10.
RF06 Ocular colobomata: clinical characteristics and systemic associations
Caroline Seghir¹, Pierre-Antoine Aymard¹, Christophe Orssaud¹, Jean-Louis Dufier¹, Olivier Roche³, Matthieu Robert¹
¹ Hôpital Necker-Enfants Malades, France

Speaker: Caroline Seghir

Introduction: Ocular colobomata (OC) may be either isolated or associated with other malformations. There is currently no consensus regarding the nature of the work-up which should be performed in a child presenting with an apparently isolated OC. The aim of this study is to describe the clinical characteristics and systemic associations of the OC seen in a tertiary paediatric centre.

Methods: We retrospectively reviewed the cases of children who presented in our institution with an OC between 1st of January 2010 and 1st of January 2013. Children with morning glory syndrome, optic disc pit or papillo-renal syndrome were not included. For each eye, the nature of the coloboma was defined as followed: type I (iris coloboma without posterior pole involvement), type II (chorio-retinal coloboma without optic disc involvement), type III (chorio-retinal coloboma affecting the optic disc), type IV (optic disc coloboma).

Results: Seventy-four children (117 eyes, 63 right, 54 left) were included – 40 girls, 34 boys. Ten children (17 eyes) presented extra-ocular malformations and were referred to in order to look for an ocular abnormality (group 1); 64 children (101 eyes) were referred to for an ophthalmological diagnosis or management of an apparently isolated coloboma (group 2). The nature of the OC was: type I in 14 eyes, type II in 27 eyes, type III in 45 eyes and type IV in 31 eyes. Among children from group 2, extra-ocular malformations were discovered in 27 cases – malformations of the kidney in 6 cases, of the heart in 8 cases, of the central nervous system in 9 cases, in the ENT field in 6 cases.

Conclusion: This series underlines the frequency of systemic malformations associated with OC, even in case of an apparently isolated ocular abnormality. It is not possible from these results to predict the probability of an extra-ocular malformation from the nature of the diagnosed OC. Systemic associations were discovered in children presenting with an apparently type I OC. We propose that a work-up including at least a paediatric clinical examination and explorations of heart, kidney and brain, be systematically performed in every case, regardless of the type of OC.

See also: Poster P11.
RF07 Microstructural retinal analysis in patients with chorioretinal coloboma
Christina Gerth-Kahlert¹, Hannes Wildberger¹
¹ University of Zurich, Department of Ophthalmology, Switzerland

Speaker: Christina Gerth-Kahlert

Introduction: Chorioretinal coloboma is a congenital anomaly due to fusional defects of the optical fissure. It can be very variable: ranging from forme fruste RPE changes with normal visual function to blind microphthalmia with extended coloboma. Patients can show reduced visual acuity (VA) although the coloboma does not involve the macular region.

Methods: Spectral-domain optical coherence tomography (OCT) was used to evaluate retinal layer abnormalities in patients with chorioretinal coloboma. Peripapillary scans and horizontal line scans over the macula region and over/adjacent to the colobomatous region were performed. Automated and manual retinal layer analysis was performed and data were compared with normative data.

Results: Data of 6 patients ages 6 to 23 years with uni- or bilateral chorioretinal coloboma were analyzed so far. VA varied from 20/20 to counting fingers. All patients with full VA showed reduced foveal thickness. Some of the patients did not show a foveal depression at all. Coloboma appeared (1) ‘empty’ without retinal layers, (2) with inner retinal layers while outer retinal layers were ‘rolled in’ at the edge of the coloboma and (3) covered with retinal layers forming a ‘tent’ over it.

Conclusion: Chorioretinal coloboma are very variable in its microstructural appearance. Microstructural analysis might give inside into developmental aspects and might predict functional outcome.

See also: Poster P12.
RF08 Frequency of spontaneous pulsations of the central retinal vein in the paediatric population
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2 lst Department Ophthalmology, Aristotle University of Thessaloniki, Greece

Speaker: Anna Mourgela

Introduction: Spontaneous venous pulsations (SVPs) of the central retinal vein have been studied in adults yet not in children. Presence of SVPs is considered to be a clinical sign helping to exclude raised intracranial pressure and differentiate pseudopapilledema from true papilledema. We aimed to investigate the presence of SVPs in a normal outpatient paediatric population.

Methods: Healthy outpatients aged 3-17 years attending our tertiary hospital for squint/refractive error were examined for presence of SVPs at the optic nerve head, cup-to-disc ratio (c/d ratio), and cycloplegic refraction. Patients with diagnosed central nervous system disease or symptoms, abnormal discs and diagnosis or suspicion of glaucoma, were excluded.

Results: Ninety-four children (49 girls) with a mean age of 103.9±38.36 months were included. Bilateral presence of SVPs was recorded in 44 (46.8%), lateral presence in 21 (22.3%) and bilateral absence in 26 (27.7%) children. One one-eyed child and two children uncooperative for examination of their second eye (3.2% in total) were excluded from the counting. Mean c/d ratio was 0.2±0.15 and mean spherical equivalent was 0.37±1.75. Results were similar for right and left eyes. No statistically significant correlation of the presence of SVPs to age, spherical equivalent or c/d ratio was observed.

Conclusion: To our knowledge this is the first study providing data on the frequency of spontaneous retinal venous pulsations in the paediatric population.

See also: Poster P13.
RF09 The role of Chromatic VEP in routine paediatric testing
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Introduction: VEP responses to chromatic (red-green, R-G and blue-yellow, B-Y) stimuli (cVEP) can give the clinician additional information on child's visual pathway integrity and function and can therefore be a useful complementary tool in everyday paediatric electrophysiological testing. The purpose of this study was to investigate cVEP response characteristics during the first year of life and to collect normative data base for babies. This study also complements our previous studies on cVEP in schoolchildren (Tekavčič Pompe et al., 2006) and preschool children (Tekavčič Pompe et al., 2012).

Methods: 44 healthy babies aged 2 to 12 months were binocularly tested. cVEP were recorded to isoluminant R-G and B-Y stimulus. The stimulus was a circle composed of horizontal sinusoidal gratings with 90% chromatic contrast and spatial frequency of 2 cycles/deg. Two stimulus sizes (7° and 21°) and onset-offset mode of stimulation (ON – 300ms, OFF – 700 ms) were used. cVEP were recorded from Oz (mid occipital) position and the reference was at Fz. Waveform characteristics and its changes throughout the first year of life were studied.

Results: cVEP responses were reliably recorded in all but two youngest babies. Characteristic cVEP response consisted of negative-positive-negative complex, positive (P) wave being far more prominent than both negative waves (N1 and N2). cVEP response to larger stimulus size (21°) showed shorter latency and higher amplitude to both R-G and B-Y stimuli compared to smaller stimulus size (7°). Same was true when comparing R-G vs. B-Y stimulus: R-G responses showed higher amplitude and shorter latency than B-Y response, for both stimulus sizes. P wave showed shortening of the latency with increasing age both for R-G (R²=0.59) and B-Y (R²=0.41) 21° stimulation, whereas P wave amplitude didn't show significant changes throughout the first year of life.

Conclusions: cVEP can be reliably recorded after the age of 3 months and show significant maturational changes throughout the first year of life. In addition to normal cVEP responses, also responses in congenital colour vision deficiency and acquired colour vision deficiency after paediatric optic neuritis will be shown.

See also: Poster P14.
RF10 Goniotomy versus trabeculotomy in primary congenital glaucoma: a five-year follow-up study

Luisa Vieira¹, Rita Anjos¹, Mariana Cardoso², Cristina Ferreira¹, Ana Xavier¹, Cristina Brito¹
¹ Centro Hospitalar Lisboa Central, ² Centro Hospitalar Baixo Vouga, Portugal

Speaker: Luisa Vieira

Introduction: Primary congenital glaucoma (PCG) is a relatively rare disease but an important cause of blindness. Treatment is mainly surgical and management is challenging. The purpose of this study is to compare outcome and complications of goniotomy or trabeculotomy for the treatment of PCG.

Methods: Retrospective case series analysis of patients with primary congenital glaucoma submitted to a goniotomy or a trabeculotomy, as the initial surgical procedure within the first year of age. A postoperative vision of 20/40 or better was considered good. Surgical success was defined as an intraocular pressure (IOP) <21 mmHg with at least a 30% reduction from preoperative pressure levels, without the need of other surgical procedures, at 5-year follow-up. Complete success was defined when target IOP was reached without the use of antiglaucoma medication, and qualified success was defined when antiglaucoma drops were required to achieve this level of IOP.

Results: A total of 20 eyes (12 patients) were submitted to goniotomy and 10 eyes (6 patients) to trabeculotomy. Of eyes in the goniotomy group, 30% had good vision compared with 40% in the trabeculotomy group. High myopia was more prevalent in the trabeculotomy group (50%) compared to goniotomy (20%). Trabeculotomy demonstrated a 20% qualified and 30% unqualified success rate with 5-year follow-up, comparing to 20% qualified and 20% unqualified success rate of goniotomy. A case of significant transitory hyphema was registered in the trabeculotomy group and there were no complications in the goniotomy group.

Conclusion: In primary congenital glaucoma, trabeculotomy appears to be as safe and effective as goniotomy within five years after surgery. Our study suggests that both procedures are still valid alternatives in the surgical management of this disease.

See also: Poster P34.
RF11 Glaucoma associated to Juvenile Xantogranuloma

Alicia Serra\(^1\), Isabel Ayet\(^1\), Jesus Diaz\(^1\), Jaume Catala\(^1\), Marta Morales\(^1\)

\(^1\)Hospital Sant Joan de Déu, Spain

Speaker: Alicia Serra

**Introduction:** Juvenile xanthogranuloma (JXG) is a benign histiocytic skin disorder that typically occurs in infancy. The eye is the organ most frequently affected, with an incidence of 0.3-0.5%. Ocular involvement includes iris infiltrates, hyphema, secondary glaucoma and rarely posterior pole or orbital affection.

**Case report:** 22 days old baby, referred to our department for Congenital Glaucoma in the right eye. Clinical signs included megalocornea, corneal edema that precluded visualization of anterior segment and fundus details, and IOP 30mmHg in the RE. After some days of medical antiglaucoma treatment, examination under general anesthesia showed a yellowish-hematic infiltrate that occupied the angle and also iris and lens. Treatment with topical steroids and cycloplegia was necessary to control the disease, and 3 months later an Ahmed valve was implanted to control the glaucoma. At 10 months of age a few yellow skin nodules were detected in the thoracic region, and the biopsy confirmed the diagnosis of JXG.

**Discussion:** Prompt diagnosis of JXG is of great importance as it may lead to severe eye complications, being secondary glaucoma the most sight threatening. Glaucoma associated with iris lesions or spontaneous hyphema is strongly suspicious of JXG. To confirm the diagnosis a thorough examination of the skin is needed, searching the cutaneous lesions and biopsy. The absence of skin involvement in a given moment does not rule out the disease, they can regress spontaneously or appear later in the evolution; in those cases an aqueous aspiration or iris biopsy can help to establish the diagnosis.

See also: Poster P35.
RF12 Outcomes in children born with unilateral persistent hyperplastic primary vitreous (PHPV)
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\textsuperscript{1}Manchester Royal Eye Hospital, UK

Speaker: Frederick Burgess

\textit{Introduction}: Analysis of visual outcomes in children born with unilateral PHPV (also known as PFV) who presented to Manchester Royal Eye Hospital (MREH), UK.

\textit{Methods}: Retrospective case series of 54 eyes of 54 patients (27 male, 27 female) with PHPV seen at MREH between September 1989 and December 2009.

\textit{Results}: 23 eyes were managed surgically, 31 non-surgically. Mean follow-up was 101 months. 74\% of surgically-managed eyes achieved at least form vision (counting fingers or better) at last follow-up, compared to 60\% of non-surgically managed eyes. In the 74\% presenting before 365 days old, age at presentation did not significantly affect acuity outcome. However median age at first operation of surgically-managed eyes achieving VA of 1.0logMar or better was 46 days compared to 61.5 days in those achieving VA of worse than 1.0logMar. 70\% of eyes with anterior PHPV achieved form vision compared to 55.6\% of eyes with posterior PHPV and 50\% of eyes with combined PHPV.

\textit{Conclusion}: Unlike previous studies, our data suggests that anteriorly located PHPV is a good prognostic indicator in comparison to combined or posterior PHPV. Our data also suggests that 46 days (rather than 77 days) is the cut-off for successful surgical intervention with better VA outcomes. The anatomical nature of PHPV appears to have the largest impact on eventual visual acuity outcomes, although early intervention, where indicated, remains important.

See also: Poster P27.
**RF13 The Long-term Psychosociological Impact of Congenital Cataract**

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Speaker: Hana Leiba

*Purpose:* To evaluate associations between congenital cataract and its therapy, and between psychological and demographic characteristics in adulthood.

*Methods:* 27 individuals who had been diagnosed with congenital cataract and 25 healthy individuals, aged 18 years and older in 2011, were asked to complete a written questionnaire. The study group was analyzed according to the age at diagnosis; type of therapy (surgery or eye patching); the presence or absence of additional eye pathologies; and unilateral or bilateral disease. Measures of self-esteem, body esteem, and social esteem were assessed.

*Results:* No statistically significant differences were found between the study and control groups in any of the examined parameters. Patients with congenital cataract who had been treated with eye patching scored higher, on average, on "comfort in physical touch (N=27, 3.458±0.406 vs. 3.058±0.466, p=0.05), and lower on "body protection" (N=27, 3.583±0.577 vs. 4.171±0.383, p=0.007), both measured on the Orbach Body Investment Scale (BIS). Patients with eye pathologies in addition to congenital cataract scored lower, on average, on "body protection" (N=27, 3.794±0.516 vs. 4.250±0.405, P= 0.019). Patients who were diagnosed before the age of 1 year scored lower, indicating a higher level of self-esteem (N=27, 1.316±0.256 and 1.657±0.393, P= 0.012) and scored higher, on average, on physical esteem (N=27, 4.347±0.418 vs. 3.842±0.382, P=0.003) more of them achieved high school matriculation (N=26, P= 0.03) and academic education (N=26, P=0.006).

*Conclusions:* Long-term psychological and demographic consequences of eye patching, additional eye pathologies, and age of cataract diagnosis were found, yet not of congenital cataract per se.

See also: Poster P25.
RF14 Visual outcome and glaucoma frequency in PECARE, a pediatric cataract register

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Speaker: Kristina Tornqvist

Introduction: Since 2007 children operated on because of pediatric cataract at <8 years of age in Sweden or at one of the two operating Danish clinics have been registered in a web-based register. Since January 2012 also the second eye clinic in Denmark operating cataract in children participate in this work.

Methods: Primary data are registered at operation and follow-up data are registered at ages 1, 2, 5 and 10 years. In this paper we will give a brief overview of data on children with congenital cataract and operated on <= 3 months of age.

Results: To date data from 458 eyes are registered. A total of 169 of these are operated on <= 3 months. A total of 123 eyes of these eyes operated at an early age had been followed-up at one year of age. Of these 44 (36%) had developed glaucoma during that first year. Visual acuity measurement were available in totally 61 of these children with VA values varying between 0 and 0.35, mean 0.09. A total of 95 eyes had been followed-up at 2 years of age. During this second year another 26 eyes had developed glaucoma. VA varied between 0.01 and 0.6 (mean 0.24) in the 46 cases were values were registered. Only 20 eyes had been followed-up at the age of 5 years. In this group another 4 eyes had developed glaucoma, visual acuities were obtained in 17 eyes with VA values between 0.01 and 0.32 (mean 0.18).

Conclusion: Congenital cataract is one of the few treatable causes of visual impairment appearing in childhood. Improved surgical techniques enable better outcome. From an amblyopia point of view an operation as early as possible is desirable which on the other hand can increase the risk for glaucoma, the complication which is no doubt the most serious threat to favourable outcome. Assembled data in our register can give us guidelines to how to best treat this fairly rare but nevertheless important disorder.

See also: Poster P26.
RF15 Persistent Fetal Vasculature (PFV) associated with high myopia (PAHM) – a distinct variant of classic PFV
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Speaker: Joanna Kate Barnes

Introduction: Characteristic findings of persistent fetal vasculature (PFV) include microphthalmos and microcornea, typically associated with hyperopia. There have been only rare reports of PFV associated with high myopia in non-glaucomatous eyes. Aim: To report a distinct clinical subset of children with PFV-associated high myopia (PAHM).

Methods: Retrospective case series of all children with PFV and extreme myopia (>minus 10D) seen by the same consultant (GDH).

Results: We report three unusual cases of PFV associated with extreme myopia, ranging between -13.50D and -31.00D. In all cases, the main mechanism of myopia was increased axial length in the affected eye compared to the normal fellow eye, the difference ranging between 2.4 and 5.7mm. One case also demonstrated posterior lentiglobus (raising axial myopia of -24.50/+2.00x45 to -31D within the central area of lentiglobus). In this case, histopathology of the posterior capsule following cataract surgery demonstrated subcapsular fibrosis and posterior migration of lens epithelial cells. We hypothesise that this histological process caused concentric constriction of the posterior capsule, leading to relatively rapid development of posterior lentiglobus and progressive index myopia. One child developed delayed extreme anisometropia and resultant visual loss at 6 years of age. None had evidence of glaucoma.

Conclusion: We propose a distinct sub-group of PFV associated with extremely high myopia (PAHM†). The underlying mechanism appears to be axial, but additional index myopia due to posterior lentiglobus and posterior subcapsular lens epithelial migration on histopathology was also seen and may occur with several years delay.

See also: Poster P28.
Norrie disease. Phenotypic characteristics, genetic evaluation and preliminary results of early surgical treatment

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Introduction: Norrie disease is a blinding ocular disorder with an X-linked recessive mode of inheritance. The disease is characterized by congenital and bilateral retinal dysplasia leading to severe ocular complications such as retinal detachment, glaucoma, or phthisis bulbi. The aim of this retrospective study is to highlight the recognizable ocular phenotype of the disease, to report the results of the genetic evaluation, and to discuss both difficulties and outcomes of the surgical procedures recently performed for the last children diagnosed with the disease.

Methods: We retrospectively reviewed 17 medical records of patients clinically diagnosed with Norrie disease from 1996 through 2013. The anatomical evolution, visual outcome, and results of the genetic study are described.

Results: The cohort included 17 male patients. Of the 17 boys of the cohort, 9 had no light perception visual acuity bilaterally and several patients developed painful complications requiring an enucleation. A mutation or deletion in the NDP (Norrie Disease Pseudoglioma) gene has been confirmed in 10 patients. A surgical procedure consisting in vitrectomy with or without lensectomy was performed for 6 patients.

Discussion: Although the disease is very rare, the recognizable phenotype together with a family history consistent with an X-linked recessive mode of inheritance allow for the clinical suspicion of the disease. The diagnosis can be definitively established by identifying mutations in the NDP gene. As described in the medical literature, the disease was characterized in most of our cases by blindness present at birth or occurring in the first months of life. Several patients developed severe and painful ocular complications. A surgical procedure was performed for 6 children. Our results suggest that early surgical treatment seem to be effective to prevent or delay the anatomical and visual deterioration and may at least prevent painful ocular complications.

Conclusion: Norrie disease is a rare blinding disorder. The clinical diagnosis is based on a recognizable ocular phenotype with B scan echography associated with a family history consistent with an X-linked recessive mode of inheritance. Although early surgical procedures for Norrie disease represent a difficult challenge and requires a highly specialized ophthalmological center, our constatations concerning the outcome of surgical treatment seem to be in line with recommendations of Walsh and collaborators.

See also: Poster P29.
RF17 Retropupillary Artisan intraocular lens in young children with traumatic aphakia following penetrating eye injuries
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1 King Edward VII Hospital, Prince Charles Eye Unit, Windsor, and Royal Berkshire Hospital, Reading, United Kingdom

Speaker: Göran Darius Hildebrand

Introduction: Visual rehabilitation of children with penetrating eye injuries is challenging and often carries a poor prognosis. When capsular support is insufficient, implantation of the aphakic Artisan (Ophtec, Netherlands) iris-claw intraocular lens (IOL) in front of the iris is an accepted alternative to anterior chamber (AC) or scleral-fixated IOL. With anterior chamber lens implantation there is concern about long-term endothelial loss. Posterior iris (or retropupillary) fixation of an Artisan IOL theoretically reduces the risk of endothelial touch.

Methods: We present three consecutive children, aged 2, 4 and 4 years, who underwent repair of penetrating eye injuries with retropupillary implantation of the Artisan IOL between March 2011 and May 2012. Two underwent secondary retropupillary Artisan IOL implantation, while the third underwent primary retropupillary Artisan IOL implantation.

Results: The children obtained best corrected visual acuities of 6/7.5 to 6/9 (0.10-0.15 logMAR) at 8, 13 and 22 months of follow-up, respectively. One child experienced transient vitreous haemorrhage secondary to surgical peripheral iridotomy which cleared spontaneously. One child developed an inflammatory membrane behind the IOL following primary lens implantation and severe intolerance of eye drops, which was successfully treated by pars plana membranectomy.

Conclusion: To our knowledge, this is the first series of young children to undergo retropupillary Artisan IOL implantation for traumatic aphakia. The procedure appears safe and highly effective in restoring vision in paediatric aphakia without capsular support.

See also: Poster P30.
RF18 Intravitreal Chemotherapy – the first line of Retinoblastoma salvage eye treatment
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Speaker: Nadiya Bobrova

Purpose: To elaborate the guideline of retinoblastoma (RB) salvage eye treatment and to study its efficiency.

Introduction: Intravitreal chemotherapy (IVC) is successfully used in cases of recurrent RB with vitreal seeding [Kaneko, Suzuki, 2003; 2004]. Combination of local (IVC) & systemic (VEC-protocol) chemotherapy was proposed for primary RB salvage treatment [Bobrova, Sorochinskaya, 2009].

Methods: The guideline of RB salvage eye treatment was elaborated: 1st line – Melphalan IVC – 24 kids (34 eyes) at age 2 mo/o – 6 y/o (17,3 ± 16,2 mo); 2nd – consecutive systemic chemoreduction VEC-protocol (23 pt – 32 eyes); 3rd – focal tumor destruction: lasercoagulation (16 eyes), transpupilar thermotherapy (9), cryotherapy (4), brachytherapy (3). EBRT at lineal accelerator – 7 cases. RB stage: T1 – 5 eyes, T2 – 7, T3a – 5, T3b – 17. Multifocal growth was in 23 eyes, RB capsule break – 12, vitreal & retinal seeds – 13, anterior chamber seeds – 1, retinal detachment – 8, jukstapapillary localization – 11. Follow up 5 – 40 (17,1 ±9,1) mo.

Results: Initial tumor response was obtained in 93,7% patients after 1st combined intravitreal & systemic VEC-protocol course. Total amount of IVC added up to 94 (1 – 6 per eye) that allowed to decrease number of systemic chemoreduction courses to 4,3 ±0,3. VEC-protocol was totally excluded in 1 child with familiar bilateral RB. New tumor focuses during treatment have appeared in 13 eyes, 7 cases had progressive growth. 9 eyes in stages T3a-T3b were enucleated due to retinal detachment (4), new focuses development (2), papilla opticus involvement (2), no tumor response (1). Pathohystologically viable tumor cells were found only in 2 eyes, retinocytoma – in 1 case, in other 6 cases – total tumor necrosis, fibrosis, calcification and degenerative changes. Summary 25 eyes (73,5%) were preserved with total tumor regress on 20 of them: T1 – 4/5, T2 – 7/7, T3a – 3/5, T3b – 6/17. Treatment is proceeding in 4 T3b stage eyes and in 1 eye with jukstapapillary RB.

Conclusion: Developed guideline of RB salvage eye treatment based on primary IVC, allowed to avoid enucleation of 73,5% eyes (even with T3a-b stages, vitreal and retinal seeding) and decrease number of chemoreduction courses to 4,3 ±0,3.

See also: Poster P38.
RF19 The response of retinoblastoma to local treatment in chemotherapy lack of effectiveness
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Speaker: Evgeniya Bulgakova

Introduction: Presently chemotherapy (CT) is recognized as the first line treatment for retinoblastoma (Rbl). However, significant number of cases of CT insufficiency requires additional local treatment – cryotherapy (CRT), thermotherapy (TTT), or brachytherapy (BT). Purpose: to evaluate the results of the local treatment methods in the cases of insufficient Rbl response to CT.

Methods: A total of 101 tumors in 43 eyes of 35 children (aged from 5 to 62 months, mean 20) with insufficient response to systemic (vincristine, etoposide, and carboplatine) and/or local CT were included into this study. Monolateral Rbl was in 9 children, bilateral in 26. Eight eyes were group A, 13 eyes were group B, 13 eyes group C, 9 eyes – group D. Number of tumors per one eye was from 1 to 11, mean 2. Tumor thickness was from 0.9 to 8mm, mean 3.0mm. Sixteen children were monocular. Forty five tumors were treated with BT, 43 with Ru-106 plaques with the apical doses from 44 to 113Gy, mean 85.2Gy, 2 tumors were treated with Sr-90 plaques with the apical doses 207 and 211Gy. In 7 eyes the plaques were relocated successively to irradiate two or three tumors. Both eyes were irradiated in 4 children. Thirty two tumors were treated with TTT, 24 with CRT. The follow-up is from 3 to 60 months, mean 19 months.

Results: The Rbl regression pattern types after each of the treatment methods are evaluated. Complete or partial regression was achieved in 43 tumors (96%) after BT, in 28 tumors (88%) after TTT, in 23 tumors (96%) after CRT. After the local treatment 41 eyes (95%) were retained. One eye was enucleated because of the tumor growth after BT, one eye because of vitreous hemorrhage without histological signs of viable tumor. Of 9 tumors irradiated with Ru-106 with the apical dose not exceeded 60Gy, mean 53Gy, complete regression was achieved in 5 (55%), while of 33 tumors irradiated with the apical dose over 60Gy, mean 85.8Gy, complete regression was achieved in 29 (87%). There were 2 (4%) recurrences after BT, 4 (13%) after TTT, 1 (4%) after CRT. Complications occurred only after BT and included nonproliferative retinopathy (n=6, 14%), papillopathy (n=4, 9%), iris neovascularization (n=3, 7%), and glaucoma (n=1, 2%). There were no cases of metastases.

Conclusion: Local treatment is a necessary and effective part of retinoblastoma treatment especially in chemotherapy insufficiency and allows to preserve 95% of the eyes.

See also: Poster P39.
RF20 Macular retinoblastoma – An outcome study following primary chemotherapy alone

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Speaker: John Bladen

Introduction: Retinoblastoma is the commonest childhood ocular cancer with excellent survival prognosis, but at the expense of vision. Tumours located at the macula lead to visual impairment, as can the destructive treatment modalities used. It has been suggested that macula retinoblastoma may have a higher relapse rate versus extra-macular retinoblastoma due to their highly vascular location. This results in some centres routinely treating with primary chemotherapy followed by a local destructive thermotherapy. The aim of this study is to review the outcomes of patients with macular retinoblastoma treated with primary chemotherapy alone.

Methods: A retrospective case review of 75 patients with 111 macular retinoblastoma tumours spanning a 14-year period (1996-2010) with a minimum two-year follow-up was conducted. Tumour severity was classified using the International Classification of retinoblastoma (ICor). Macula was defined as an area within the temporal arcades. Outcomes included survival, macula relapse rate (defined as any new tumour within the original macula tumour pre-treatment area including overlying vitreous and subretina), enucleation rate, size of macula tumour and LogMAR visual acuity.

Results: The mean age of diagnosis was 9 months old (range 0.25-33 months). Of the 75 patients recruited 34 were female and 41 were male. Twenty-eight patients had de-novo mutations, 28 were inherited, and 19 had unknown genetic aetiology. The mean follow-up time was 89.6 months (range 48-132 months). Bilateral tumours were found in 50 individuals, the remaining 25 patients had unilateral disease. The mode ICor classification was D (35%) with B being the second most frequent (30%). ICor A=8%, B=30%, C=14%, D=35%, E=14%. Outcomes: survival 100%; macula relapse rate 23%; enucleation 25%; size of tumour range 1-30mm; and mean final visual acuity post-treatment 0.52 (range 0.02 to 3.00).

Conclusion: Primary chemotherapy alone confers an acceptable macular relapse rate of macular retinoblastoma over a long follow-up period. Our study shows similar outcomes to those centres that employ primary chemotherapy with additional thermotherapy. Visual outcome is variable and is dependent on proximity of the tumour to the fovea.

See also: Poster P42.
RF21  Macular retinoblastoma & visual function: beyond visual acuity

Pablo Durán-Pérez¹, Jaume Català-Mora¹, Margarita Sanz-Arboix¹, María Costa-González², Jesús Díaz-Cascajosa¹, Joan Prat-Bartomeu¹
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Speaker: Pablo Durán-Pérez

**Introduction:** Retinoblastoma management is devoted to life sparing conservation of the eye with the best affordable vision.

**Methods:** Clinical review of 68 eyes with retinoblastoma from 47 patients. We have included 20 eyes from 17 patients with tumor margin or scars involving the foveola, < 3 mm from optic disc or macular subretinal fluid. We have done a full clinical & ophthalmological evaluation with distance & near visual acuity, colour testing, biomicroscopy & retinography. Visual abilities like walking, detecting different objects and colours have been recorded in every infant.

**Results:** Visual acuity ranged from LP to 20/40. Visual function was not correlated to distance visual acuity. Patients with only one eye even with massive macular involvement tend to manage better than expected & usually show impressive visual abilities. Patients with asymmetric disease tend to have worse visual function in the macular or optic nerve affected eye than the first group of patients.

**Conclusion:** Evaluation of visual function and not only visual acuity should be mandatory in these patients. New amblyopia treatment modalities could be considered in patients with asymmetric disease regarding the possible benefits of gaining visual function.

See also: Poster P43.
RF22 Choroidal osteoma: rare familial occurrence
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Speaker: Mary van Schooneveld

Introduction: choroidal osteoma is usually seen unilaterally in young healthy females. Familial occurrence has been rarely observed.

Methods: we present the long-term follow-up of bilateral choroidal osteomas in a sister and a brother and a possible form fruste in the mother. Recently, bilateral submacular osteomas were discovered with ophthalmoscopy at the age of 3-months in the baby girl of the young female patient.

Conclusion: choroidal osteomas may lead to severe visual impairment as was the case in our family and may occur bilaterally with autosomal dominant inheritance.

See also: Poster P48.
Introduction: The purpose of this study was to analyze the pathological classification of the pediatric ocular tumors.

Methods: At the Department of Ophthalmology, Semmelweis University from January 2009 to April 2013 161 cases of pediatric ocular tumors (below the age of 18 years) were reviewed retrospectively. The pathological classification was done according to the localisation.

Results: The analysis covered 85 benign tumors of the eyelids. The two most frequent tumors were cysts (36 lids: 28 dermoid, 8 epidermoid cysts) and nevi (15 eyes). There were 52 eyes with conjunctival and corneal tumors, all were benign tumors. The most common tumor was conjunctival nevus (34 eyes). There were 10 eyes with benign tumors of the orbit, 9 cases of cysts and 1 case of lymphangiectasia. There were 13 eyes with histopathologically confirmed malignant intraocular tumors. All of these cases were diagnosed as retinoblastomas.

Conclusion: In the reviewed cases most of the pediatric tumors were benign and congenital. The most frequent malignant intraocular tumors were retinoblastomas. In the reviewed period we had more cases of retinoblastomas, but only 13 required enucleation.

See also: Poster P49.
RF24 Congenital nasolacrimal duct obstruction: a definitive approach to management of a common condition
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Introduction: Conflicting reports remain in the literature regarding a definitive approach to management of congenital nasolacrimal duct obstruction (CNLDO). We sought to clarify the most appropriate treatment regimen.

Methods: We conducted a retrospective analysis of 177 patients undergoing probing +/- intubation to treat CNDLO in a single institution (Royal Victoria Hospital, Belfast) from 2006-2011.

Results: 245 eyes were included in this study: 186 (76%) eyes had successful outcome at first intervention with primary probing while 56 (23%) underwent secondary intervention. There were no significant differences by gender, age or obstruction complexity between the successful and unsuccessful patients with first intervention. For those patients requiring secondary intervention, 20/30 (67%) eyes had successful probing while 24/26 (92%) had successful intubation. Patients with intubation as a secondary procedure were significantly more likely to have a successful outcome (P=0.025).

Conclusion: Primary probing for CNLDO has a high success rate. Findings suggest that if initial probing is unsuccessful then nasolacrimal intubation rather than repeat probing yields a significantly higher success.

See also: Poster P57.
RF25 An unusual evolution of preseptal cellulitis to subperiostean abscess
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Nose, Throat Department, MITERA Children's Hospital, ³ Department of Paediatric Imaging, MITERA Children's Hospital

Speaker: Athanasios Fitsios

Introduction: A 4,5 years old boy was examined for eyelid and periorbital erythema and oedema of the left eye.

Methods: The clinical ophthalmological examination confirmed the diagnosis of preseptal cellulitis as no other orbital cellulitis signs or symptoms were present. No conjunctivitis was present and not skin trauma or insect bites were found. The child was admitted to the hospital and intavenous broad spectrum antibiotics were administered intravenous. Only after 3 days the eyelid erythema seemed to diminish significantly. The fourth day a new sign was present: the left eye presented displacement of the globe (dystopia) and small proptosis. We decided to proceed to otorinolaringological (ENT) examination and imaging investigation (MRI).

Results: Otorinolaringological endoscopic examination found purulent secretions in the middle meatus of the nose, which is an indirect sign of sinusitis (ethmoidal or frontal). The imaging investigation (MRI) confirmed the ethmoidal sinusitis and discovered a subperiostean abscess. We decided not to proceed to surgical operation (functional endoscopic sinus surgery, FESS) and to cure the small patient with daily nasal drainage, decongestants and change the intravenous antibiotics. The next imaging procedure (MRI) that took place after 5 days shown quite complete resolution of the abscess and was not necessary surgical intervention.

Conclusion: When a child presents a preseptal cellulitis without skin trauma and with white eye (without ocular surface inflammation) we must pay more attention in the evolution of the clinical signs, we must think other causes that bring to that clinical entity and evaluate if it's worth to submit the child promptly to endoscopic examination of the nose.

See also: Poster P58.
RF26 Transocular orbitotomy: a new approach for optic nerve tumors in children
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1 Hospital Sant Joan de Deu de Barcelona

Introduction: Optic nerve tumors are usually removed through a cranectomy because orbital approaches are difficult in these cases. We propose to remove the pediatric orbital optic nerve tumors in blind eyes through a new surgical technique named transocular approach (with previous enucleation).

Methods: One meningioma and two optic nerve glioma NF1 negatives in small children (1, 3 and years old) had been operated on because a deforming proptosis in blind eyes because of optic nerve tumors. The technique is as followed: enucleation, dissection of the tumor from orbital tussues from the anterior created approach, removing of the optic nerve tumor cutting at the bottom of the orbit while the surgeon apply traction and, finally, an 20 mm orbital implant is sutured to the muscles. So, there are two surgeries in one. Results and complications are analized after this kind of surgery.

Results: All the surgeries could be performed completely, no orbital relapse has been seen after a 3 years of follow-up. Motility of the implant was normal in one case and limited horizontally in other. One patient had blepharoptosis that was compensated by the artificial eye.

Conclusion: Transocular orbitotomy is an interesting approach in optic nerve tumor surgery with proptotic and blind eyes in children. It should be considered as a good alternative to a craniotomy in same cases.

See also: Poster P54.
RF27 Deep periocular infantile capillary haemangiomas responding to topical timolol maleate
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Speaker: Kanmin Xue

Introduction: Periocular infantile capillary haemangiomas (IH) can cause visual impairment as well as facial disfigurement. Oral propranolol is increasingly used as first-line therapy for IH. While propanolol is usually well tolerated by infants, adverse reactions could occur in around 7% of cases. A growing body of evidence suggests that the topical β-blocker, timolol maleate, is also effective in the treatment of superficial IH but with fewer adverse effects. Almost all published reports so far have focused on topical timolol treatment of superficial IH. We presents two infants with deep periocular IH treated with topical timolol alone.

Methods: A retrospective review of two infants with deep periocular capillary haemangiomas. Case 1 was a 7-week-old with a deep IH involving the whole left upper lid, causing lid closure, astigmatism and amblyopia. Case 2 was a 6-month-old with a 15 x 20 mm medial right lower lid IH, causing limitation of adduction in depression of the right eye. Both were treated with topical timolol maleate 0.5% gel 3 drops applied twice daily to the skin.

Results: In both cases, topical timolol treatment led to near complete involution of the deep capillary haemangiomas with functional recovery of vision and satisfactory cosmesis. Case 1 received 1 year of topical timolol treatment and achieved equal visual acuity of 6/7.5 in each eye. No evidence of recurrence was seen after cessation of treatment for 5 months at the last follow-up. Case 2 was treated for 11 months and achieved visual acuity of 6/9 in each eye with full range of extraocular movements. No adverse reaction was seen.

Conclusion: These cases represent novel examples of the usage of topical timolol maleate 0.5% gel in the treatment of deep periocular capillary haemangiomas and suggest that topical timolol may be a viable alternative when systemic propranolol treatment is contraindicated.

See also: Poster P52.
RF28 Active surgical management in infantile capillary hemangiomas of the orbit and eyelids
Nadiya Bobrova\(^1\), Svitlana Tronina\(^1\)
\(^1\) SI Filatov Institute of Eye Diseases and Tissue Therapy NAMS of Ukraine

Speaker: Svitlana Tronina

Introduction: Capillary hemangioma is the most common vascular tumor in babies and infants, which can be considered as a disembioplastic neoplasm or hamartoma. The pathology characterized by different forms, clinical course and growth rate.

Purpose: To analyze the efficacy of surgical management of different clinical forms of capillary hemangiomas of the orbit and eyelids in infants.

Methods: 243 children aged 1-16 months (mean age 5 ± 2.9 months) were operated on at the Pediatric ophthalmopathology department of the SI “The Filatov Institute of Eye Diseases and Tissue Therapy of NAMS of Ukraine”. Rapid growth of the lesion was evident and documented confirmed in all patients. In superficial forms of hemangioma active progression clinically resulted in forming of typical strawberry nevus with affecting of extensive skin areas. Deep forms produced amblyogenic conditions – eyelids deformation, eye fissure narrowing with visual axis occlusion and high risk of obscurative amblyopia development. The combined staged method included cryodestruction of superficial intradermal lesions and surgical removal of the subcutaneous and/or orbital part of the tumor in different combinations depending on the form and depth of hemangioma spreading was applied.

Results: Usage of cryodestruction usually on the first stage of treatment allowed to achieve gentle scarring of the skin. Surgical excision of the deep part of the tumor eliminated disfigurement and eye occlusion avoiding amblyopia development. Good cosmetic and anatomic result was achieved in 90.4% of cases.

Conclusion: The main indication for active surgical treatment of progressive capillary hemangiomas is fast enlarging of the lesion with involving of large area of eyelids and periorcular area, significant deformity of eyelids with narrowing of eye fissure. In due time started treatment in cases of superficial hemangiomas give the possibility to avoid extensive skin area affection and reduce the time of treatment. In deep and combined forms surgical methods allow to perform radical tumor excision, achieve good cosmetic and functional result in short term and quickly remove amblyogenic factor.

See also: Poster P53.
RF29 A clinical classification system for Paravenous Pigmentary Chorioretinal Atrophy (PPCRA) in Childhood
Göran Darius Hildebrand¹, Dorothy Thompson², Isabelle Russell-Eggitt², David Taylor²
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Speaker: Göran Darius Hildebrand

Introduction: PPCRA is often mistaken for retinitis pigmentosa. No review of PPCRA among children has been undertaken so far.

Methods: Review of all known cases of PPCRA under the age of 18 years in the world literature.

Results: We identified 27 cases of paediatric PPCRA, including 3 new cases. We identified two distinctive patterns of presentation: Primary PPCRA (22/27, 81%) presenting with PPCRA, gradual visual loss, frequent positive family history (12/22, 55%) and no clear triggering event. Secondary PPCRA (5/27, 19%) presenting with sudden, profound, simultaneous bilateral visual loss due to bilateral neuroretinitis (3/5) or acute maculopathy (2/5). PPCRA phenotype developed during the resolution of the acute episode. Secondary PPCRA was non-familial and associated with measles-related neuroretinitis (2 cases), post-vaccination neuroretinitis (1 case) and acute maculopathy (two cases).

Conclusion: We present a simple clinical classification system for paediatric PPCRA. Secondary PPCRA may be the longterm phenotypic outcome of neuroretinitis and acute maculopathies in childhood.

See also: Poster P62.
RF30 Choroidal neovascularization secondary to Autosomal Recessive Bestrophinopathy in a 9-year-old girl treated by intravitreal ranibizumab injection
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Speaker: Marta Pawlak

Introduction: Authors present the case of 9-year-old girl with choroidal neovascularization secondary to Autosomal Recessive Bestrophinopathy treated by intravitreal ranibizumab injection.

Methods: The Autosomal Recessive Bestrophinopathy was diagnosed based on results of ophthalmological and electrophysiological examinations. It was confirmed by genetical testing – two BEST1 mutations were found (c.74G>A and c.55C>T). A year after the diagnose was established, choroidal neovascularization in patient’s left eye was diagnosed. It was treated with one intravitreal ranibizumab injection.

Results: During one year observation period improvement in left’s eye visual acuity was observed.

Conclusion: Intravitreal ranibizumab injection is an effective therapeutic option in Autosomal Recessive Bestrophinopathy.

See also: Poster P63.
RF31 Identical Twins Exposed! – Bestrophinopathy, Autosomal Recessive?
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Speaker: Lintje Ho

Introduction: Best disease is generally perceived as a Mendelian macular dystrophy, transmitted as a dominant trait caused by mutations in the BEST1 gene, but also autosomal recessive inheritance has been described. We report of a case of unusual expression of best-like disease in one sibling of a twin-pair.

Methods: Clinical features of the proband were documented by means of best-corrected visual acuity (BCVA), color fundus photographs, fundus autofluorescence (FAF), optical coherence tomography (OCT), electro-oculogram (EOG), electroretinogram (ERG), and a visual field test (Humphrey 10-2). His twin brother and his mother underwent a FAF, OCT and EOG. Blood samples of both twins were taken for genetic sequencing of the BEST1 gene.

Results: The proband had a Snellen BCVA of 0.7 and 0.6. Color fundus photographs and FAF of the proband showed a diffuse irregularity of the reflex from the retinal pigment epithelium, including dispersed punctate flecks. There was a very low EOG light rise, and a reduced paracentral ERG. Photographs and FAF for his twin brother and mother were normal. The OCT for all three family members showed no abnormalities. DNA sequencing revealed that the proband had two heterozygous missense mutations in the BEST1 gene: c.679T>A (p.(Tyr227Asn)) and c.934G>A (p.(Asp312Asn)). His twin brother was a carrier of only one missense mutation: c.670T>A(p.(Tyr227Asn)). This missense mutation was inherited from the father, and has been described as an autosomal dominant mutation (Petruhkin et al 1998). Conclusion: Two heterozygous mutations in the BEST1 gene lead to an early, atypical manifestation of Best disease in a seven-year old. Surprisingly, the difference in BEST1 genotype revealed that the presumed monozygotic twins were not identical! Moreover, the difference in fundus phenotype between the twins suggests that Best disease in this case may represent an autosomal recessive form. Alternatively, clinical manifestations may still occur later in life. The variable expression of Best disease remains unexplained. Other genes in addition to BEST1 and/or environmental influences may play a role in the wide range of clinical expression seen. Further research to correlate specific mutations and candidate (genetic) modifiers with various phenotypic aspects (such as severity and age of onset) may be warranted.

See also: Poster P64.
RF32 Megalopapilla in children: definition by OCT
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Speaker: Elena Jarrín Hernández

Introduction: The term megalopapilla was coined for enlarged optic disc not associated with any other morphological anomalies. By definition, the diameter of the optic nerve head has to be greater than 2.1 mm. Its detection is important because it may be associated with encephalocele. In healthy children, the enlarged cup associated to megalopapilla may be confused with infantile glaucoma. Our aim is to characterize megalopapilla by optical coherence tomography (OCT) to update its definition employing this current ancillary test.

Methods: Healthy children, whose optic nerve heads had a diameter greater than 2.1 mm manually measured using the caliper of the digital retinography, were recruited. All of them underwent a complete ocular examination, including intraocular pressure by aplanation, biomicroscopy of the anterior and posterior poles and optical coherence tomography. Optic nerve head and macular data obtained were compared with a control group.

Results: Seven children were included; six with bilateral megalopapillas. Three were male. Three were caucasians, three hispanic and one from the south of Asia. Any of them showed anomalies like posterior embriotoxon in the anterior pole biomicroscopy and their intraocular pressure was under 18 mmHg in all cases. The optic disc area measured by OCT ranged from 2.21 to 3.19 mm3 (mean 2.64 mm3, SD 0.27). Mean average cup/disc ration was 0.7 (SD 0,39), ranging from 0.66 to 0.79. Mean retinal nerve fiber layer (94.77 μm), temporal RNFL (70.92 μm), and ganglion cell layer (83 μm) thicknesses and mean macular volume (10.04 mm3) were not significantly different from mean values of healthy children.

Conclusion: Optical coherence tomography may be helpful when evaluating children with large optic nerve cups. A mean optic disc area above 2.2 mm3 and normal RNFL and macular measures may lead to the diagnosis of megalopapilla, automatically measured by OCT. Average cup/disc ratio greater than 0.8 might be pathological.

See also: Poster P66.
RF33 Laser flare photometry in healthy preschool children

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Speaker: Judit Körtvélyes

Introduction: In adult patients normal aqueous flare is increasing with age, but there is no data in healthy preschool children at all. Our aim was to measure aqueous flare with laser flare meter in preschool children.

Methods: The measurements were performed with Kowa Laser Flare FM-600 device. It is an objective, non invasive, non contact method for measuring aqueous flare level. We examined one eye of 50 healthy preschool children lacking any ocular diseases. Average age was 5.94 years, (range 3-7 years), 25 girls and 25 boys. While the measurements took place the children were seated in a nurse’s lap.

Results: Average flare value was 3.53, SD: 1.76. The highest value was 12.1 and the lowest was 1.1.

Conclusion: This average value was lower than it was measured in young healthy adults before. The examination itself required a few minute of concentration to stay nice and still. At this age, not all the children are capable to do so. Children with disorganised movements, body schema disturbance, vestibular disorder, pencil grip problem, find it very difficult to concentrate steadily, therefore failing to produce accurate measurements and hence some of them can not even be tested at all.

See also: Poster P65.
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POSTERS
P01  Orbital, Preseptal and Facial Neurofibromatosis
Eren Cerman¹, Muhsin Eraslan¹, Begum Dirican¹
¹ Marmara University School of Medicine, Istanbul, Turkey

Introduction: A neurofibromatosis case with preseptal mass was presented to discuss the best approach to this unusual presentation.

Methods: A 3.5 year old boy was referred to our clinic due to type 1 neurofibromatosis, ptosis and a hemifacial lesion. The patient underwent orbital magnetic resonance (MR) imaging and punch biopsy was performed on upper tarsal area.

Results: On physical examination he had a hemifacial cutaneous skin-colored lesion. The lesion was including left periorbital region, malar area and upper lip. On ophthalmologic examination the patient had a mild restriction in upper and lateral gaze. He had proptosis and 2 mm ptosis. MR imaging revealed a subcutaneous and preseptal lesion with irregular borders, that was extending into the lateral orbital area and lateral orbital wall involvement was apparent. Pathologic investigation of the lesion revealed a plexiform neurofibroma.

Conclusion: A neurofibromatosis related cutaneous lesion might be removable but subcutaneous lesions may cause disfigurement, and may extend into the orbita and cause extraocular muscle disorders. To choose the best treatment modality for such a lesion is difficult.

P02  Functional visual loss or brain tumour? The use of sweep VEP
Maria van Genderen¹, Ymkje Hettinga¹, Frans Riemslag¹, Gerard de Wit¹
¹ Bartiméus Institute for the Visually Impaired, the Netherlands

See: Rapid Fire RF01.
P03  Macrocephaly and Papilloedema: Neurofibromatosis Debut?
Elena Jarrín Hernández¹, Laura Cabrejas Martínez², Susana Noval Martín³, Ana Isabel Pastor Vivas¹, Javier García Gil De Bernabé¹, María Esther Arranz Márquez¹
¹ Rey Juan Carlos Hospital, Mostoles (Madrid); ² Fundación Jiménez Díaz Hospital (Madrid); ³ La Paz Hospital (Madrid)

Introduction: The eye is a window to suspect many important pathologies. The ophthalmologists must be alert to the signs of possible syndromes, especially in children.

Methods: Case: A 7-year-old child was evaluated at the Emergency Department because of a cranial traumatism for an accidental fall suffering a transient consciousness loss. His family and personal medical history were unremarkable. On examination a macrocephaly with regular morphology was observed, as well as motor coordination clumsiness, mild dysarthria, frequent vomiting, drowsiness and a café au lait macule in one leg. His parents had detected some mild neurologic disturbances before the fall. The Computed Tomography revealed a chronic triventricular dilation and a marked thinning of the white matter and the corpus callosum. The aqueduct was severely stenosed by an expansive mass in the quadrigeminal tubercula. There was another mass in the fourth ventricle floor. These lesions seemed to be low grade glial tumors or hamartomas. On ophthalmic examination visual acuity was 1 in both eyes. Extrinsic and intrinsic ocular motility were normal. Slit-lamp examination showed Lisch nodules. An image of chronic papilloedema was observed in the funduscopy.

Results: He was referred to neurosurgery and endoscopic ventriculostomy (ETV) was performed. The cerebrospinal fluid output pressure was very high. In the postoperative the child is much more alert, with no tendency to drowsiness. Papilloedema is resolving ophthalmoscopically and by OCT. NF1 genetic test has been applied.

Conclusion: Neurofibromatosis is a tumoral disease caused by a change in chromosome 17. Half of the cases are de novo mutations. Glial tumors are common in the CNS. The endoscopic ventriculostomy is the procedure of choice for hydrocephalus associated with neurofibromatosis. The failure of ETV can lead to patient death so close monitoring is essential.
P04  Ophthalmological findings in different types of pediatric CNS tumors
Ulrika Lidén¹, Agneta Rydberg¹, Falkman Lena¹, Ingrid Axelsson¹, Kerstin Hellgren¹
¹ Pediatric neurooftalmological clinic, Sweden

Introduction: Presenting early ophthalmological symptoms, long term sequels and rehabilitation strategies in a cohort of pediatric CNS tumors at a pediatric university hospital.

Methods: During a three year period we consecutively included all children referred to the pediatric neuroophthalmological clinic at Astrid Lindgren's children's hospital with newly diagnosed CNS tumors. A thorough clinical examination including visual fields, ocular motility and fundus imaging was done by the authors.

Results: A group of 27 children with brain tumors were included in the project. At the time of referral, ophthalmological findings were common, various and often previously missed in this group of children. During follow up, ophthalmological findings were often used for timing of tumor treatment. Different rehabilitation strategies were applied, depending on the symptoms.

Conclusion: The ophthalmologist plays an important role in detecting early symptoms leading to early diagnosis and treatment in many types of pediatric CNS tumors. The role in rehabilitation of residual visual problems and detecting worsening of visual status is equally important.
P05  **The visual outcome of the 16-year old patient post cerebellum surgery**  
Marta Michalczuk¹, Beata Urban¹, Alina Bakunowicz-Łazarczyk¹  
¹ Medical University of Bialystok

**Objectives:** The operations of brain tumors that affect visual pathway cause specific visual field changes or even overall blindness. Those complications are a direct outcome of the mechanical removal of the neoplastic brain tissue located within the visual pathway, or vicariously the result of its damage during tumor surgery in the nearby localization. However, the surgery of the central nervous system neoplasm, with no correlation to the optic pathway, can also cause changes in visual function.  

**Methods:** 16-year-old boy, who underwent surgery of the right half of the cerebellum on the grounds of astrocitoma pilocitico, was examined in the Clinic of Ophthalmology. Before and after the brain operation patient underwent fundoscopy, visual acuity test, CT and MRI scans of the brain. Additionally, OCT of the optic disc, autorefractometry, colour vision tests, visual field examination and PVEP were performed only after the brain operation.  

**Results:** The patient’s visual acuity was reduced in both eyes just 8 months after the brain surgery and at present amounts to 5/8 in the right eye to 5/7 in the left eye. The colour vision examination, refraction test and fundoscopy revealed no abnormalities. The OCT of the optic disc presented the asymmetry of global RNFL thickness between eyes (thicker in every segment of the right eye), which still was normal in both eyes. Perimetry revealed changes in the visual field. PVEPs showed increased latencies of the P100 wave RE>LE during stimulation with 15 minutes pattern. Before and after operation CT and MRI scans of the brain showed no visual pathway involvement.  

**Conclusions:**  
1. The fluctuations of intracranial pressure and oxidation stress during brain tumor surgery should always be considered as a possible cause of visual dysfunction.  
2. All patients after any brain surgery should be constantly and acutely examined by an ophthalmologist.

P06  **Optic pathway gliomas in children and adolescents with or without Neurofibromatosis type 1**  
Kamilla Rothe Nissen¹, Jens Folke Kiilgaard¹  
¹ Department of Ophthalmology University Hospital Rigshospital Copenhagen, Denmark

**Introduction:** Optic pathway gliomas in children with Neurofibromatosis type 1. In current practice, the goal of visual assessments in children with optic gliomas is to identify, non refractive abnormalities in visual function. The majority of gliomas are identified prior to the age of 6 years, during the period of visual maturation. A stagnation or deterioration in vision can be difficult to detect. Visual progression, defined as a two-line decrement in visual acuity, is an indication for chemotherapy. Therefore initially management involves close observation of the visual acuity.  

**Methods, Results, Conclusion:** We have a evolved a database in ACCESS, monitoring visual progression in graphics clarifying the visual deterioration.
P07  Efficacy and safety of a combination of 0.5% Cyclopentolate with 2.5% Phenylephrine used for ROP screening in 623 consecutive examinations spanning a 30 month period
James Neffendorf¹, P. Michael Mota¹, G. Darius Hildebrand¹
¹ Royal Berkshire Hospital, Reading and King Edward VII Hospital, Windsor, UK

See: Rapid Fire RF02.

P08  Long term refractive status follow-up of prematures: singletons vs multiples
Hana Leiba¹, Niv Levy², Eric Shinwell¹
¹ Kaplan medical center , Rehovot, Israel, ² Hadassah Medical School, the Hebrew University, Jerusalem, Israel

See: Rapid Fire RF03.

P09  Macular function measured with mfERG in prematurely-born children at school-age
Hanna Åkerblom¹, Gerd Holmström¹
¹ Dep. of neuroscience, Uppsala University, Sweden

See: Rapid Fire RF04.

P10  Twins with bilateral posterior microphthalmos
Yvette Braaksma-Besselink¹, Marije Sminia¹, Linda Groenveld¹
¹ Academic Medical Centre (AMC) Amsterdam, the Netherlands

See: Rapid Fire RF05.

P11  Ocular colobomata: clinical characteristics and systemic associations
Caroline Seghir¹, Pierre-Antoine Aymard¹, Christophe Orsaud¹, Jean-Louis Dufier¹, Olivier Roche¹, Matthieu Robert¹
¹ Hôpital Necker-Enfants Malades, France

See: Rapid Fire RF06.
P12  Microstructural retinal analysis in patients with chorioretinal coloboma
Christina Gerth-Kahlert¹, Hannes Wildberger¹
¹ University of Zurich, Department of Ophthalmology, Switzerland

See: Rapid Fire RF07.

P13  Frequency of spontaneous pulsations of the central retinal vein in the paediatric population
Anna Mourgela¹, Nikolaos Ziakas², Stavros Dimitrakos¹, Asimina Mataftsi¹
¹ IInd Department Ophthalmology, Aristotle University of Thessaloniki, Greece,
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See: Rapid Fire RF08.

P14  The role of Chromatic VEP in routine paediatric testing
Manca Tekavčič Pompe¹, Branka Stirn Kranjc¹, Jelka Brecelj¹
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See: Rapid Fire RF09.

P15  Adalimumab and Refractory Uveitis in Children
Ana Clement¹, Isabel Valls¹, Maria Luisa Andres¹, Clara Jimenez¹
¹ Hospital Universitario Niño Jesus, Spain

Introduction: To study the efficacy of Adalimumab in pediatric uveitis refractory to treatment with Methotrexate.

Methods: We retrospectively evaluated 38 pediatric patients with uveitis unresponsive to methotrexate who were treated with Adalimumab at a dose of 40mg/m² every 2 weeks for a mean of 31.9 months. Twenty-eight patients with juvenile idiopathic arthritis, 7 with idiopathic anterior uveitis, 2 idiopathic panuveitis and one Blau syndrome associated uveitis.

Results: The number of flares per year decreased from 4.41 to 0.98. Visual acuity remained stable in 33 patients, improving in 3 due to the resolution of a previous macular edema and in 2 after cataract removal. There were 3 cases of ocular hypertension that required topical treatment. Adalimumab had to be suspended in 2 patients due to anaphylactic reactions. No other serious systemic adverse events were observed. Adalimumab was removed in 12 patients (after a mean of 39.9m), 11 relapsed, first relapse occurring at a mean of 3.3m. In 6 patients Adalimumab was reintroduced, five of them remaining stable.

Conclusion: Adalimumab appear to be a useful and safe therapy for children with uveitis resistant to treatment with Methotrexate.
Introduction: A grandmother, her son and her grandson have developed a chronic bilateral neuritis accompanied by severe macular edema. The disease appears in advance in the next generation. Infections and environmental etiologies have been excluded. We present this complex familial case to share our doubts and to know the experts opinions.

Methods: The father was diagnosed of bilateral neuritis when he was 8 years old and he received no treatment. He suffered an important decrease of visual acuity in the twenties with any other symptoms. His mother is nearly blind because of neuritis and retinitis pigmentosa. She has macular and optic nerve atrophy with a mild pigment dispersion surrounding the macula. In the man’s funduscopy we observed a severe neuroretinitis with a great and bilateral optic disc swelling, vasculitis and one granulomatous tumor. MRI, lumbar puncture and the systemic work-up was negative. He has been treated with acetazolamide, oral, intravitreal and intravenous steroids with no success. He received empiric antibiotics for Bartonella. In the angiofluoresceingraphy, ischemic was observed in the periphery. Suspecting IRVAN syndrome, he received photocoagulation of ischemic areas. Intravitreous Bevacizumab was injected with no response. His visual acuity is 0,1 in both eyes. His 3-year-old son has a bilateral and asymmetric optic nerve swelling with a macular star in the worst eye. Suspecting IRVAN syndrome after negative MRI, work-up and lumbar puncture, he has received some doses of intravenous immunoglobulins with a great anatomic response but the swelling increases when the effect finishes after one month. Metrexato was associated getting some stability. His visual acuity is 0,1 in the right eye with the macular edema and 0,8 in the left eye (E Snellen).

Results: Suspecting a genetic disturbance in axoplasmic transport OPA gen has been studied in the family but no mutations have been found. Finally, we think this rare entity may be an ocular variant of Chronic Infantile Neurological Cutaneous and Articular/Neonatal Onset Multisystem Inflammatory Syndrome (CINCA/NOMID). The father is being treated with interleukin-1 receptor antagonist (Anakinra). He has not experienced visual fluctuations with the treatment and the macular edema is thinner.

Conclusions: This is a challenging pathology because of its severity, the familial association, the absence of known diagnosis and the difficult response to treatments.
Introduction: Uveitis of childhood is a very serious and potentially blinding disease. Chronic recurrent course of the disease, high frequency of complications, frequent combination of infectious and non-infectious immunopathological mechanisms significantly complicate the management uveitis in children. The purpose of the study was to increase the efficiency of chronic recurrent uveitis management in children by improvement of the diagnostic algorithm and development of the differential etiopathogenetic therapy.

Methods: There were observed 52 children with chronic recurrent uveitis aged from 5 to 18. Serological tests with the study of the specific antibodies IgG, IgA, IgM levels against Herpes simplex virus (HSV), Cytomegalovirus (CMV), Epstein-Barr virus (EBV), Chlamydia trachomatis, Toxoplasma Gondii of serum and polymerase chain reaction (PCR) with the definition of the pathogen DNA were performed. The combination therapy of the uveitis in children caused by herpetic infection in the case of replicative activity of the infections consists of the systemic acyclovir 1000 mg and recombinant IFN alpha-2β 1mln per day during 3-4 weeks. Treatment with fansidar 2tablets per week during 6 weeks was applied for patients with uveitis caused by Toxoplasma Gondii. Azithromycin therapy was prescribed in cases of Chlamydia etiology uveitis. Methylprednisolone 8-12 mg per day in combination with azathioprine 50 mg per day was used adjuvant in treatment for the uveitis in children in case of presence of autoimmune component.

Results: All patients have chronic recurrent severe course of uveitis. All patients were diagnosed with the persistent intracellular infections, including herpetic infection in 36 (69%) children, herpetic and toxoplasmosis co-infection in 12 (23%) patients and chlamydial infection was detected in 4 (8%) cases. Replicative activity with DNA of pathogen detection was observed in 18 (34,6%) patients, including DNA of EBV in 10 (19%), DNA of CMV in 6 (11,5%) and DNA of HSV in 2 (4%) patients. In all patients, combination therapy was reported to obtain immediate and long-term positive clinical outcome. The control PCR investigation for 4-6 weeks revealed the absence of the pathogen DNA.

Conclusion: Proposed and clinically approved the diagnostic algorithm and etiopathogenetic therapy of the chronic recurrent children uveitis caused by persistent intracellular infections provides immediate and long-term clinical and laboratory remission of the disease.
P18  Identifying genes for cerebral visual impairment
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Cerebral visual impairment (CVI) is one of the major causes of visual impairment in the developed world, as it accounts for 27% of low vision in childhood\textsuperscript{(1)}. It includes all visual dysfunctions caused by damage to, or malfunctioning of, the retrochiasmatic pathways in the absence of any major ocular disease\textsuperscript{(2)}. CVI can consist of a reduced visual acuity, and/or visual field defects. Furthermore, there can be an abnormal visual behavior, such as staring into light or delayed fixation. Deficits in higher perceptual functions, for example difficulties with recognition of objects and faces, or visuospatial disorders can occur, and are sometimes the only features of CVI\textsuperscript{(3)}. CVI is often part of a more complex phenotype, consisting of intellectual disability, epilepsy and/or deafness. An important cause of CVI is acquired damage to the brain, mainly the result of perinatal problems, such as cerebral hemorrhage or periventricular leucomalacia. However, in about 40% of the patients the underlying cause is unknown, but significant evidence for genetic causes have been reported. We included 60 patients with CVI and a visual acuity \(\leq 0.3\) without any known cause for the CVI. All patients were investigated by a pediatric ophthalmologist and a clinical geneticist. To identify genetic defects for CVI, we tested 10 patients and their unaffected parents (trio approach), in which causal copy number variation were previously excluded, using exome sequencing. In exome sequencing the coding regions of more than 21,000 genes are sequenced for possible pathogenic mutations. The data were analyzed for X-linked, autosomal recessive and de novo autosomal dominant causes. We identified several candidate genes for CVI in our patients which will be presented.

\textbf{Acknowledgements:} This study was accomplished in part through the Centers for Mendelian Genomics research effort funded by the National Institutes of Health and supported by the National Human Genome Research Institute grant U54HG006542 to the Baylor-Hopkins Center for Mendelian Genomics. This work has been supported by grants from Stichting ODAS that contributed through UitZicht and the Vereniging Bartiméus-Sonneheerdt (5781251).

P19 Molecular analysis and ocular manifestations of Coffin Siris Syndrome
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Introduction: Coffin Siris syndrome (CSS) was first described in 1970 by Coffin and Siris; it is a rare genetic disorder with fewer than 100 individuals reported to date. It is clinically heterogeneous, but characterized by the presence of the following 3 features (i) fifth digit nail/distal phalanx hypoplasia/aplasia, (ii) developmental delay and (iii) coarse facial features including wide mouth with thick, everted upper and lower lips, broad nasal bridge with bulbous upturned nasal tip, and hypertrichosis of the eyebrows and eyelashes. This study describes the ocular manifestations of CSS, which has not previously been comprehensively documented in the literature, and to investigate the genetic causality.

Methods: Two unrelated patients with CSS were recruited: patient I is a 13 year old girl; and patient II is a 3 year old boy. Full history and ophthalmic examination was undertaken. Peripheral blood samples were collected and genomic DNA extracted. All 20 exons (including intron/exon boundaries) of ARID1B gene on chromosome 6, known to cause one-third of CSS cases, were amplified using standard PCR protocol, followed by automated sequencing. For the detection of deletions/duplications multiplex ligation-dependent probe amplification (MLPA) was performed.

Results: Both patients had reduced visual acuity. Patient I had a best corrected visual acuity (BCVA) of 0.80 in the right eye (RE) and 0.50 in the left eye (LE) using LogMAR. She was myopic (RE -5.00DS/-1.00DCx80 and LE -5.50DS/-1.00DCx80), with a right exotropia, and right anomalous optic disc. Patient II had a BCVA 6/38 with both eyes open (using Cardiff cards at 50 cm), was emmetropic with a manifest oblique jerky nystagmus, left ptosis, intermittent left esotropia, and right anomalous optic disc. Electrophysiology revealed macular pathway dysfunction on pattern VEPs. Both patients had mutations in the ARID1B gene: Patient I had a heterozygous de novo frameshift mutation c.1259insA (p.Asn420fs) in exon 1; and Patient II had a heterozygous de novo deletion involving exons 6-9.

Conclusion: This study has characterized the range of ocular features associated with CSS and identified different genetic mutations in the disease-causing gene ARID1B. It highlights the requirement for regular ophthalmology review of patients with CSS to ensure visual outcomes are optimized.
P20  Case report of Acute Anisocoria in a 20-month-old child

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Introduction: A 20-month-old boy was sent for ophthalmologic examination due to acute anisocoria (presented 2 hours before) referred from his mother.

Methods: In the opthalmological examination not signs for eyelid ptosis and extraocular muscle palsy were present. From the observation of the pupils we found that the right pupil was normal in size and in responsiveness to light, while the left pupil was dilated and with only a minimum reaction in light and near reflex evaluation. Slit lamp (biomicroscopic) examination shown no sphinter damage or other anterior segment findings. Fundus examination was within normal limits. There was no history of recent trauma and the neurologic and paediatric examination remained without other findings. No other symptoms were present like headache, ocular pain or altered consciousness.

The last 3 days the child was treated with ipratropium bromide inhalation aerosol. We decided not to proceed to further imaging (CT, MRI) or invasive (lumbal puncture) examination and to re-examine clinically the child in 24 hours.

Results: Both pupils after 24 hours were equally normal in size and in light reaction. In the next 7 days examination, the pupils showed symmetrical and normally reactive. Inadvertent ocular exposure to ipratropium (anticholinergic drug), caused by ill-fitting face mask, seemed a plausible explanation of the abnormal dilated pupil.

Conclusion: An abnormal dilated pupil could be alarming because may be an early sign of neurologic emergency. Ipratropium bromide should be considered in the differential diagnosis of patients, especially in paediatric patients, because of the difficulty of maintaining proper face fit during respiratory treatments.

P21  Visual abnormalities in adopted children and adolescents

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Introduction: The aim of the study was to evaluate ophthalmological findings in adopted children.

Methods: The study was conducted from 2011 to 2013. Children born before 32 weeks of gestation or with birth weight less than 1500 g were excluded from the study. Thirty seven children and adolescents (mean age 8.3 years; range 4-16 years; 18 boys) were included in the study. All patients had detailed ophthalmological examination. Additionally, pattern visual evoked potentials was also performed in each child.

Results: Thirty six patients were referred to ophthalmologist because of ophthalmological complaints (strabismus, nystagmus, tearing, eyes rubbing) and one – to rule out ophthalmological abnormalities. Authors describe ophthalmological and electrophysiological findings.

Conclusion: Authors want to highlight the importance of early ophthalmological examination in this group of patients. Unknown family history and high probability of adverse prenatal events oblige to detailed ophthalmological and, sometimes, neurological evaluation. The early diagnosis allows early intervention and revalidation to optimize children’s outcome.
P22  A rare Alström Syndrome case with remarkably well preserved visual function
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Introduction: Alström syndrome is an autosomal recessive ciliopathy engaging multiple organs and causing early profound visual loss. We describe a rare case of this syndrome with well-preserved visual function in a longitudinal study.

Methods: A Swedish patient diagnosed with Alström syndrome has been followed from infancy to age 18 years at the Pediatrics, Ophthalmology and Audiology Departments at Lund and Örebro University Hospitals. Full somatic status, audiology, as well as the visual function have been carefully monitored. The visual function has been repeatedly documented with visual acuity, visual fields, electroretinogram, fundus pictures and optical coherence tomography.

Results: This patient who has otherwise the characteristic features of Alström syndrome with obesity, sensorineural loss and cardiomyopathy shows a markedly well preserved visual function, despite the long duration of the disease, with visual acuity over +1.0 log MAR, visual fields with normal peripheral but affected central isopters, cone dysfunction documented by ERG and pigmentary changes of the retina.

Conclusion: This patient is unique in Sweden and one of the few subjects with Alström syndrome known globally to be visually functional beyond adolescence. This could be attributed to genetic or environmental factors.

P23 Congenital fixed dilated pupils: from the pupil into multisystemic disorders
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Congenital fixed dilated pupils, sometimes referred as congenital mydriasis or partial aniridia is characterized by aplasia or hypoplasia of the iris muscles, with absence of iris between collarette and pupillary border, and consequently a scalloped pupillary margin. Although iris morphology is pathognomonic, it has sometimes been misdiagnosed as aniridia. Congenital mydriasis is the key point for diagnosis of the newly described ACTA2 mutation. It has also been described in Gillespie syndrome, or in Megacystis-microcolon-intestinal hypoperistaltis syndrome (MMIHS). Careful diagnosis must exclude pharmacologic mydriasis, bilateral compressive III nerve palsy, or bilateral tonic pupils and autonomic neuropathies. Fixed dilated pupils in a young child is an extremely rare condition whose presence should alert paediatricians and ophthalmologists to the possibility of the co-existence of systemic life-threatening disorders.
P24 Preliminary results of the macular and retinal nerve fibre layer thickness, measured with Cirrus and Stratus OCT in full-term children
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Introduction: The primary aim was to investigate the macular thickness and retinal nerve fibre layer, assessed with Cirrus optical coherence tomography (OCT), and to report the repeatability and reproducibility. A second aim was to compare Cirrus and Stratus OCT.

Methods: Twenty-five children, 5-15 years of age, born at term with normal birth weights, randomly chosen from the birth register of the Swedish National Board of Health and Social Welfare, were examined. Three measurements of the macula and the optic nerve head were done by one person, and thereafter repeated by another examiner. Finally, three measurements were performed with the Stratus OCT. Visual acuity, refraction in cycloplegia and a fundus examination were also performed.

Results: The macular thickness was noted in all nine ETDRS-areas (A1-9) with Cirrus and Stratus OCT. Measurements with Stratus OCT revealed thinner maculae than with the Cirrus OCT. The mean central macular thickness (A1) in the right eye (RE) was 253 μm (range 224 – 280) and in the left eye (LE) 255 μm (range 220 – 282), when measured with Cirrus OCT. The mean central macular thickness (A1), measured with Stratus OCT, was 201 μm (range 181 – 232) and 207 μm (range 190 – 286) in RE and LE's, respectively. Regarding the optic nerve head, the mean average retinal thickness was 100 μm (range 75 – 123) in both eyes, the disc area was 1.7 mm² (range 0.9-2.8) in RE’s and 1.9 (range 0.9-2.9) in LE’s and the rim area was 1.4 mm² (range 0.9-2.2) in both eyes, assessed with Cirrus OCT. Finally, repeatability and reproducibility of the Cirrus OCT will be reported.

Conclusion: There are no normative data available for the macular thickness and the retinal nerve fibre layer assessed with Cirrus OCT in children. Such data are of importance when used as a comparison to children with diseases of the macula and the optic nerve. The values assessed with Cirrus and Stratus OCT differed and the measurements cannot be interchanged.

P25 The Long-term Psychosociological Impact of Congenital Cataract
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See: Rapid Fire RF13.
P26 Visual outcome and glaucoma frequency in PECARE, a pediatric cataract register
Kristina Tornqvist¹, Birgitte Haargard², Gunilla Magnusson³, Alf Nyström³, Annika Rosensvärd⁴
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See: Rapid Fire RF14.

P27 Outcomes in children born with unilateral persistent hyperplastic primary vitreous (PHPV)
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¹ Manchester Royal Eye Hospital, UK

See: Rapid Fire RF12.

P28 Persistent Fetal Vasculature (PFV) associated with high myopia (PAHM) – a distinct variant of classic PFV
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See: Rapid Fire RF15.

P29 Norrie disease. Phenotypic characteristics, genetic evaluation and preliminary results of early surgical treatment
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See: Rapid Fire RF16.
P30 Retropupillary Artisan intraocular lens in young children with traumatic aphakia following penetrating eye injuries
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¹ King Edward VII Hospital, Prince Charles Eye Unit, Windsor, and Royal Berkshire Hospital, Reading, United Kingdom

See: Rapid Fire RF17.

P31 Congenital Cataract in a Child with Pyridoxine-Dependent Epilepsy
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Introduction: Pyridoxine-dependent epilepsy is an autosomal recessive cause of neonatal epileptic encephalopathy with an incidence of 1:20,000 to 1:687,000 live births. Ophthalmic disease has never been documented in association with pyridoxine dependent epilepsy.

Methods: We describe the novel observation of a child noted to have a right-sided congenital cataract with an intractable neonatal epileptic encephalopathy. At 1 year of age, extensive biochemical investigation identified pyridoxine-dependent epilepsy as the cause of her seizures – abolished with oral pyridoxine monotherapy. Aged 5 years, she was noted to have progressive, bilateral, asymmetric cataracts.

Results: Pyridoxine-dependent epilepsy is the result of mutations in the ALDH7A1 gene encoding Antiquitin, an enzyme protective against cellular dehydration and osmotic stress. Accumulating metabolic precursors in PDE have been demonstrated strongly cataractogenic in vitro. Experimental pyridoxine deficiency in swine has been associated with lenticular opacities in vivo. The association of ALDH7A1 haploinsufficiency in pyridoxine-dependent epilepsy and congenital cataract may offer novel insight into the relationship between osmotic stress and foetal cataract development. Bilateral progression of cataracts in this child suggests ongoing metabolic dysregulation within the crystalline lens despite pyridoxine supplementation at doses sufficient to control seizures.

Conclusions: We have identified a potential relationship between pyridoxine dependent epilepsy and congenital cataract. The probability of co-incidental pyridoxine-dependent epilepsy and congenital cataract in the same child is between 1 in 80 million and 1 in 2 billion. Further clinical reports are required to establish a clear relationship between PDE and congenital cataract. We suggest ophthalmic examination of children with pyridoxine-dependent epilepsy to exclude lenticular opacities.
Black Magic in Pseudophakic Eyes: Ultra-Widefield Retinal Imaging through Black IOLs for Intractable Diplopia

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Background: Black intraocular lens implantation is a treatment option in patients with intractable diplopia. Despite high rates of post-operative satisfaction, their clinical use has been limited by concerns that an inability to visualize the retina through Black IOLs may prevent the diagnosis of important posterior pole diseases, principally malignant melanoma. We successfully acquired SLO/OCT images in a patient with an implanted Black IOL. We present our body of novel research identifying the range of retinal imaging capabilities in this patient group.

Methods: Spectral transmission profiles of all clinically available Blacks IOLs (Morcher, Artisan iris-claw and Dr Schmidt’s) were defined using broad-spectrum and near-infrared (NIR) light sources and a spectroradiometer. We implanted a model eye with each occlusive IOL and acquired retinal profiles using seven commercially available SLO/OCT imaging systems (Heidelberg, Opko, Optovue, Topcon, Canon, Zeiss, Bioptigen). We acquired widefield retinal cSLO images (835nm) using the Staurenghi contact lens and a non-contact ultra-widefield SLO imaging system (Heidelberg Engineering) to determine the extent of peripheral retinal imaging possible through a Black IOL in a calibrated model eye. We evaluated the utility of near-infrared transillumination photography to image the anterior uveal tract through Black IOLs.

Results: Spectral transmission profiles identified the presence of a NIR window of high-level transmission across Morcher Black IOLs. Black IOLs produced by other manufacturers did not transmit NIR light. All clinically available SLO/OCT scanners identified subtle retinal features of a model eye through Morcher Black IOLs. Contact and non-contact cSLO widefield imaging systems were able to image the retinal periphery to 150 degrees through a Black IOL. NIR transillumination photography may identify anterior uveal melanomas in patients with Black IOLs.

Conclusion: This research has identified the first sensitive retinal imaging modalities to detect retinal disease in current patients with Black IOLs at follow-up. SLO/OCT, ultra-widefield cSLO and NIR transillumination photography are able to detect melanoma at any anatomical location in the uveal tract in patients with Black IOLs. This research may permit more patients to benefit from Black IOL implantation as the inability to image the retina should no longer influence clinical judgement.
P33  Anterior segment developmental anomaly in a twin with the foetal transfusion syndrome
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Introduction: This presentation aims to describe a rare case of bilateral congenital corneal opacity (CCO) secondary to kerato-irido-lenticular dysgenesis in one of identical twins who suffered the foetal transfusion syndrome.

Methods: Case report.

Results: A prematurely born (gestational age 33 weeks, birth weight 1640 gr) girl, one of monozygotic twins, was noted to have CCO at birth. The other twin had normal eyes. At week 20 of the pregnancy the mother had laser occlusion for anomalous transplacental shunt and twin-twin transfusion syndrome. Imaging of the anterior segment showed kerato-irido-lenticular adhesions in the left eye, irido-corneal adhesions in the right eye and intra-corneal cystic space in both eyes. B scan ultrasonography of the posterior segment showed no abnormalities and the corneal size and intraocular pressure revealed no signs of glaucoma as yet. The infant was programmed for corneal transplantation in the left eye and a large iridectomy in the right eye. Genetic testing is pending.

Conclusion: In this rare occurrence of CCO in a twin with the foetal transfusion syndrome, where the identical twin is normal, genetic testing will shed some light on the role of intrauterine ischemia in anterior segment developmental anomaly.

P34  Goniotomy versus trabeculotomy in primary congenital glaucoma: a five-year follow-up study
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1 Centro Hospitalar Lisboa Central, 2 Centro Hospitalar Baixo Vouga, Portugal

See: Rapid Fire RF10.

P35  Glaucoma associated to Juvenile Xantogranuloma
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1 Hospital Sant Joan de Déu, Spain

See: Rapid Fire RF11.
P36 Visual acuity outcome in congenital glaucoma
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Introduction: Congenital glaucoma is a rare condition often associated with significant visual loss. The purpose of this study is to evaluate the visual acuity outcome in paediatric glaucoma.

Methods: Retrospective review of patients older than 3 years with congenital glaucoma surveilled in our institution. Clinical presentation, medical and surgical therapy, visual outcomes and refractive errors were recorded.

Results: A total of 27 eyes from 16 patients with a mean follow up of 100 (46-156) months were included in our study. A visual acuity > 20/40 was achieved in 10 patients. Presentation at birth, bilateral disease, delay of surgical treatment and multiple procedures were associated with poor vision (< 20/200). Myopia was found in 70% of patients, with high myopia being frequent.

Conclusions: While congenital glaucoma may result in poor vision, a good visual outcome was achieved in most of our patients. There is a clear tendency to a myopic shift in eyes affected by this disease.

P37 Congenital Glaucoma in Greece: An 11-Year retrospective study
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Introduction: To evaluate the management of all primary congenital glaucoma cases seen at our units serving the population of northern and southern Greece.

Methods: A retrospective study was carried out on all new congenital glaucoma patients who were managed at the department of pediatric ophthalmology of the Aristotle University of Thessaloniki and A.Kyriakou Children’s Hospital in Athens during the period 2002-2013.

Results: A total number of 30 patients with primary congenital glaucoma presented to our clinics during the last 11 years. 21 were boys and 9 girls. 21 of them had bilateral glaucoma and 9 unilateral. 47 of 51 eyes were operated. Of those 12 had goniotomy, 6 goniotomy and trabeculotomy, 10 goniotomy, trabeculotomy and trabeculectomy with antimetabolite, 6 goniotomy and trabeculectomy, 6 trabeculotomy, 3 trabeculectomy and 4 valve insertion. Average age at presentation was 22 months and follow-up range 1-110 months. Complications noted were phthisis in 4 eyes, cataract in 8 eyes, which were successfully surgically managed in 6. Control of IOP was achieved in 40 of 47 not phthisical eyes, of which 15 required the use of additional topical medications.

Conclusion: This study provides useful information regarding the incidence and management of congenital glaucoma in Greece. Pediatric glaucoma must be timely detected and treated promptly.
P38 Intravitreal Chemotherapy – the first line of Retinoblastoma salvage eye treatment
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See: Rapid Fire RF18.

P39 The response of retinoblastoma to local treatment in chemotherapy lack of effectiveness
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See: Rapid Fire RF19.
P40  New Concepts in Chemotherapy for Retinoblastoma

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Introduction: Systemic chemotherapy has been the standard treatment for retinoblastoma since the 1990s. It is not, however, without side-effects, both short-term (such as myelosuppression) and long-term (such as infertility and secondary leukemias). It has also proved to be poorly effective for vitreous and retinal seeding. New concepts in chemotherapy have been developed over the last decade. We present the preliminary results of three of these: super-selective intra-arterial melphalan; periocular injection of topotecan and intravitreal injection of melphalan.

Methods: Super-selective intra-arterial chemotherapy is carried out by catheterism of the ophthalmic artery via the femoral artery, under fluoroscopy. This enables direct delivery of the chemical agent to the ophthalmic artery. Periocular topotecan involves injection of the chemical agent directly into the posterior sub-Tenon space, using a 25 G needle. Correct distribution of the drug around the globe is assessed by ultrasound after the procedure. Intravitreal injections of melphalan are carried out using a 32 G needle. An anterior chamber puncture of aqueous liquid of the same quantity is performed to prevent vitreal reflux of fluid contaminated with potentially malignant cells. Upon needle removal three cycles of freeze and thaw cryocoagulation are applied to sterilize the needle track.

Results: 94 eyes of 86 children suffering from advanced or recurrent, severely pre-treated retinoblastoma, received ocular chemotherapy delivered via the ophthalmic artery or by intravitreal or periocular injection, or a combination thereof. The remission rate is over 90% for each of the categories. Only 6 eyes required enucleation due to disease progression. The most frequent ocular complications were: choroidal occlusive vasculopathy for intra-arterial drug delivery, salt and pepper retinopathy at the injection site for intravitreal injections, and mild transitory myelodysplasia for periocular injections.

Conclusion: New delivery modes of chemotherapy enable tumor control in eyes previously destined for external beam radiotherapy or enucleation. The higher intraocular drug concentration for a lower dosage also reduces secondary systemic effects. The encouraging results of this preliminary study justify the multicentric phase II clinical trial set up by the Swiss Pediatric Oncology Group (SPOG_RB_2011), to better define indications, evaluate response and determine any complications.
P41 Alternating intra-arterial and intravitreal chemotherapy for advanced intraocular retinoblastoma: first successful results without systemic chemotherapy
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Background: To describe the efficacy of intra-arterial chemotherapy combined with intravitreal chemotherapy for the treatment of advanced stage retinoblastoma.
Methods: the Authors report the medical records of four patients who presented with unilateral retinoblastoma, (Reese-Ellsworth stage Vb/ group D of ABC Classification in the affected eye). They underwent clinical and ophthalmoscopic evaluation and MRI to exclude local and CNS dissemination. All 4 patients received 2 cycles (six infusions) of intrarterial chemotherapy (4-5 mg of melphalan/0.3-0.4 mg of topotecan), and from 7 up to 9 melphalan injections into the vitreous (10-30 μg in 0.05 mL).
Results: Successful control of tumor masses and vitreous seeds was achieved in all cases at long-term (> 6 months) follow-up. Tumor control was 100% in all cases. Complications included: posterior lens opacities, acute ischemic papillitis, transient hypotonia, vitreous hemorrhage. There were no cases of orbital tumor recurrence or metastasis (follow-up range, 6 – 12 months).
Conclusions: Sequential intra-arterial chemotherapy and intravitreal melphalan for advanced retinoblastoma allowed to provide retinal and vitreous seed control.

P42 Macular retinoblastoma – An outcome study following primary chemotherapy alone
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See: Rapid Fire RF20.

P43 Macular retinoblastoma & visual function: beyond visual acuity
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See: Rapid Fire RF21.
P44  Changes in the management of retinoblastoma in the central region of Hungary
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Introduction: Our aim was to analyze and compare the patients data who were diagnosed and treated because of retinoblastoma at the Department of Ophthalmology in Semmelweis University in two periods (January 2003 – December 2007 and January 2008 – March 2013).

Methods: A retrospective study was performed. Retinoblastoma was diagnosed in 12 children (6 girls and 6 boys) in the first period and 20 children (15 girls and 5 boys) in the second period. The mean age was 15.6 and 17.2 months at the diagnosis. Family history was positiv in 2 and 4 cases. Bilateral form was observed in 6 and 7 children.

Results: The wide-field digital photography was used for documentation from 2010 (17 cases). Telemedical screening in neonatal units of our University leads to detection of one case. We could save 8 eyes in the first period and 11 eyes in the second period with intravenous chemotherapy and local tretament. Enucleation was performed in 9 children (10 eyes) during the first period and in 13 children (14 eyes) during the second period. Genetic examination was performed in cooperation with University of Duisburg-Essen in 12 children.

Conclusion: We could improve the management of the retinoblastoma in our unit with photodocumentation, genetic testing and treatment modalities which target to save the eye with cooperation other centres.

P45  The case early diagnosis of Retinoblastoma with RetCam
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¹ North-West State Medical University

5 mln. people live in St. Petersburg (child population ~ 800 000). Usually 4 – 5 cases of retinoblastoma are registered in St. Petersburg per year. Case report: A boy was born in St. Petersburg on September 2010. He was the second of twinges (extracorporeal fertilization, EF). His birth weight was 1350 g, gestational age – 29 weeks. He was being examined by ophthalmologist as a premature baby with RetCam. The type I of ROP was diagnosed when he was 36 weeks of PA. He was treated by diode laser (wave length – 532 nm). After 2 weeks of the laser coagulation the boy was examined again. The ophthalmologist saw the signs of the regress of ROP. At that photo has been made with RetCam III now the observer can see a little white spot is located in the paramacular zone. But at that time nobody thought about retinoblastoma. At the next photo (after 2 weeks) focus of the tumor was observed clearly and it contained of all macular zone. Since that time (3 month y.o.) the patient has been treated in Germany (Essen). The boy received 6 courses of the chemical therapy. After 2 courses of the chemical therapy the stabilization of changes has been observed. But after 1.5 months new nodules of the tumor appeared. Selective a.ophthalmica perfusion of Melphalan was made. When the boy was 1 year old, in spite of the treatment, the eye has been enucleated, but the other eye is healthy now. This case confirms the opportunity of the RetCam for the early diagnosis of retinoblastoma.
P46  Syndromic Retinal Dystrophy associated with Astrocytic Hamartoma of the Optic Nerve Head
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Introduction: The association of optic disc astrocytic hamartoma with retinal dystrophy is rare. Most of the astrocytic retinal hamartomas are pathognomonic for phakomatoses like tuberous sclerosis.

Methods: A 14 year old Caucasian boy with suspected retinitis pigmentosa was referred to our department for further diagnostic work up. In the pediatric examination truncal obesity and mild intellectual disability was documented. The ophthalmic assessment included visual acuity testing, funduscopy, optical coherence tomography (OCT), autofluorescence imaging, full-field and pattern ERGs.

Results: The patient showed a reduced visual acuity on both eyes. The undetectable pattern ERGs indicated severe macular dysfunction, the undetectable dark and light adapted full-field ERGs indicated severe generalized dysfunction of the rod and cone system in keeping with the diagnosis of a rod-cone-dystrophy involving the macula. The fundus examination showed peripheral irregular pigmentation, but the really impressive finding were bilateral tumors on the optic nerve head looking like astrocytic hamartomas typical for tuberous sclerosis remaining unchanged during follow-up. By a comprehensive screening (cranial MRT, abdominal ultrasound, echocardiography, dermatologic examination) no other signs for tuberous sclerosis could be found. No mutation, duplication or deletion was identified in TSC1 and TSC2. Based on the clinical findings a syndromic retinopathy was suspected and we initiated a comprehensive molecular genetic screening for Bardet Biedl or similar syndromic retinopathies.

Conclusion: Astrocytic hamartomas typical for tuberous sclerosis can be found in retinal dystrophies. The differential diagnosis in syndromic cases is even more difficult.

P47  3 D Optical coherence tomography in retinoblastoma: residual tumor, calcificate and chorioretinal scar after eye-saving therapy
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¹ Moscow Helmholtz Research Institute of Eye Diseases

Introduction: To reveal diagnostic symptoms of residual retinoblastoma, calcificate and chorioretinal scar after eye-saving therapy, documented by 3 D optical coherence tomography.

Methods: The analysis of retinal changes at 21 children (21 eyes, 27 focuses) with retinoblastoma after chemotherapy and local treatment (brachytherapy and transpupillitary thermotherapy): residual tumor – 12, chorioretinal scar – 12, calcified focus – 3, revealed by 3 D optical coherence tomography, was carried out.

Results: Tomographical symptoms of chorioretinal scar: flat bright hyperreflective stripe replacing all retinal layers. Calcified focus: raised bright thinning hyperreflective layer under which the effect of «shadow». Residual tumors: elevation, homogeneity, hyperreflectivity of tumor structure and thickening of the tomographycal scan, equal choroidal profile and on the periphery – bright flat hyperreflective retina (scar).

Conclusion: 3D optical coherence tomography allows to diagnose residual retinoblastoma, calcificate, chorioretinal scar after eye-saving.
P48  **Choroidal osteoma: rare familial occurrence**
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See: Rapid Fire RF22.

P49  **Pathological analysis of pediatric ocular tumors**
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¹ Department of Ophthalmology, Semmelweis University Budapest, Hungary

See: Rapid Fire RF23.

P50  **Complications following enucleation with primary orbital implant in retinoblastoma**
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*Introduction:* Assess complications following enucleation with primary orbital implant in children with retinoblastoma from September 1999 to September 2012 in our hospital.

*Results:* A total of 50 surgeries were included with a minimum follow up of 6 months. 22 eyes were primarily enucleated, 8 eyes were enucleated following chemotherapy and 20 eyes were enucleated following chemotherapy and external radiotherapy. Major complications included implant exposure (4 eyes), cellulitis (1 eye) and anophthalmic socket syndrome (7 eyes). 9 out of 10 eyes with these complications had previously received external radiotherapy. We encountered only 3.6% of major complications following primary enucleation or chemotherapy and enucleation in comparison with 45% when enucleation was preceded by chemotherapy and external radiotherapy.

*Conclusions:* External radiotherapy is the major risk factor for severe complications following enucleation with primary implant in our series.

P51  **withdrawn**
P52  Deep periocular infantile capillary haemangiomas responding to topical timolol maleate
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See: Rapid Fire RF27.

P53  Active surgical management in infantile capillary hemangiomas of the orbit and eyelids
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See: Rapid Fire RF28.

P54  Transocular orbitotomy: a new approach for optic nerve tumors in children
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¹ Hospital Sant Joan de Deu de Barcelona

See: Rapid Fire RF26.
P55 Glomuvenous malformation of the orbit – report of three cases
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Introduction: Glomuvenous malformations (GVM) are characterised by cutaneous lesions consisting of distended vascular channels with abnormal glomus cells. The glomulin gene is located on chromosome 1p21,22. Here we present 3 cases of GVM involving multiple structures of the eye and orbit.

Methods: Retrospective chart review of clinical data, course and investigations (Doppler orbital ultrasound, MRI, histopathology, and genetic studies) was performed.

Results: Three females aged 1-7 (now with 2-5 year follow up) presented with multiple soft bluish-pink cutaneous, intermittently painful lesions. These gradually enlarged from birth and involved eyelids, tarsal conjunctiva, optic nerve sheath and extraocular muscles in all cases. Other affected sites included the anterior chest wall, axilla, back, leg, face and vulva. Photographs of clinical findings, Doppler orbital ultrasound, MRI, histopathology, genetic studies and treatment results will be presented. No patient has developed compromised vision at this stage. One patient underwent partial debulking of upper eyelid lesion.

Conclusion: The literature reports one orbital glomangioma, two cases with solitary glomangiomas of the extraocular muscles, one case with conjunctiva involvement, three solitary glomus tumors of the eyelids, and four cases with involvement of the eyelid in plaque-like glomangiomatosis. Our cases show an unusual presentation of GVM involving the orbits, eyelids, extraocular muscles and conjunctiva. GVM mimics common venous malformations because of the blue hue. However, with experience, it can be clinically and pathologically distinguished, which may subsequently influence multidisciplinary team management. Patients with periocular and facial GVM should be reviewed by an Ophthalmologist to exclude any orbital involvement and to manage possible orbital complications.
P56  A diagnostic challenge: severe orbital Kimura disease in a Caucasian child
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**Background:** Kimura disease (KD) is a rare chronic inflammatory disorder characterized by angiolymphatic proliferation and eosinophilic infiltration. This condition, which predominantly affects young men of Asian origin, is of unknown etiology.

**Purpose:** To report a case of orbital KD in a non-Asian child.

**Design:** Interventional case report.

**Case:** A 12-year-old Caucasian boy presented with a painless mass in his right upper eyelid, accompanied by ptosis, progressive exophthalmos, reduced eye motility, sicca symptoms, diminished visual acuity, and transient photopsia in the right eye. CT and MRI scans of the right orbit revealed diffuse swelling of the upper eyelid with a distinct nodular mass, enlargement of the lacrimal gland, as well as enlargement of the medial, superior, and lateral recti, and superior oblique muscles, causing proptosis and segmental optic nerve compression along the optic nerve canal.

**Investigations and Management:** Excisional biopsy from the skin lesions along with markedly elevated serum IgE levels and eosinophilia confirmed the diagnosis of KD. Periorbital triamcinolone acetonide and iv methylprednisolone pulse therapy were initiated. There was a prompt clinical improvement and no complications were encountered.

**Conclusion:** To our knowledge, this is the first non-Asian case of pediatric orbital KD presenting with impending compressive optic neuropathy.

P57  Congenital nasolacrimal duct obstruction: a definitive approach to management of a common condition
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See: Rapid Fire RF24.

P58  An unusual evolution of preseptal cellulitis to subperiostean abscess
Athanasios Fitsios¹, Efthymios Kalampalikis², Georgia Papaioannou³, Dionisios Nikolopoulos¹, Eleonora Marouli¹
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See: Rapid Fire RF25.
Progressive conjunctival nevi cryodestruction in pediatric patients

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Introduction: Conjunctival nevus is common benign lesion in children and teenagers. Among different tumors of conjunctiva it frequency amounts to 25,6 %.

Purpose: To analyze the results of progressive conjunctival nevi treatment with application of cryodestruction in pediatric patients.

Methods: Cryodestruction of progressive conjunctival nevi were performed in 273 children and teenagers (275 eyes) aged 3-17 (mean age 10,14 ± 3,27) y/o. Nonmelanocytic vascular nevi were observed in 101 children (102 eyes – 37,1 %), pigmented – in 172 children (173 eyes – 62,9 %). Perilimbal localization was more typical for nonmelanocytic nevi. Pigmentary nevi localized on the perilimbal bulbar conjunctiva (61,7 %), lacrimal caruncle (29,0 %), plica semilunaris (8,4%) and tarsal conjunctiva (0,9%). Fluorescent angiography was included to the complex pre-op examination for real size of the lesion determination. Contact dosed cryodestruction using special tip with liquid nitrogen as a refrigerant providing temperature – 130ºC on the working surface was applied in all cases. The time of cryodestruction exposition during the session and number of sessions varied depending on the clinical features – prominence and area of the lesion, intensity of pigmentation. In different cases from 1 up to 4 sessions of cryodestruction (average 1,66 ± 0,8) was performed. The number of cryosurgeries averaged 1,56 ± 0,81 sessions in nonmelanocytic lesions and 1,68 ± 0,75 in pigmentary nevi.

Results: Early term after cryodestruction characterized by slight inflammatory reaction of lesion and surrounding area, which completely reduced after 2-3 weeks. The remote results of treatment was observed in terms from 3 months till 5 years (average 19,7 ± 12,5 months). Complete resorption of the nevus – both a vascular component and a pigment, was achieved in all cases with no scarring or other change of relief and structure of tissues at the locus of criodestruction.

Conclusion: Fluorescent angiography is a highly informative method for pre-op diagnostics of conjunctival nevi and post-op monitoring of the treatment results. Dosed cryodestruction is the high-performance method of progressive conjunctival nevi treatment in pediatric patients allowing to achieve complete lesion resorption of any localization and size that makes it adequate alternative to surgical excision.
P60 Exophthalmos and Growth Hormone Replacement Therapy
Ceren Gurez¹, Eren Cerman², Muhsin Eraslan², Volkan Dericioglu²
¹ Yalova State Hospital, Yalova, Turkey, ² Marmara University School of Medicine, Istanbul, Turkey

Introduction: Exophthalmos is rare in pediatric population. A case of pediatric exophthalmia was presented and the role of growth hormone therapy in exophthalmos was discussed in the light of literature.

Methods: A 9 year old girl with Turner syndrome who had proptosis since 1 year was referred for further investigation. Screening for metabolic diseases, magnetic resonance imaging and genetic analysis was performed.

Results: The genotype analysis of the patient revealed XO Turner syndrome. She had normal thyroid function. She was using growth hormone (GH) replacement since 2 years. On ophthalmic examination the visual acuity was 20/20 bilaterally. Slit lamp and fundus examination was normal. One year follow up revealed a developing exophthalmos and lid lag.

Conclusion: Though exophthalmos due to GH treatment is not reported before and there is no direct evidence that in this patient GH is causing exophthalmos, GH is known to induce exophthalmos and may be related in this case.

P61 Severe pediatric Graves orbitopathy in adolescents of African origin
Andrea Papp¹, Clemence Vasserot-Merle², Guido Dorner¹, Dion Paridaens²
¹ Department of Pediatric Ophthalmology and Strabismus, Department of Ophthalmology and Optometry, Medical University of Vienna, Austria, ² Department of Oculoplastic and Orbital Surgery, The Rotterdam Eye Hospital, Rotterdam, the Netherlands

Purpose: To report on two cases of severe pediatric Graves orbitopathy (GO) in two adolescents of African origin.

Design: Interventional, retrospective case report.

Methods: Two black male adolescents presented with highly active GO and signs of beginning compressive optic neuropathy. None of them were smokers or had a family history of GO. Besides urgent referral to pediatric endocrinologists, weekly iv methylprednisolon pulse therapy was initiated.

Results: In spite of the fluctuating thyroid hormone levels in the initial phase of antithyroid therapy, iv. steroid administration stopped the progression of malignant GO rapidly in both of our patients without any considerable side effects.

Conclusion: Although the course of GO during childhood is considered to be mild, severe, sight threatening GO – requiring immunosuppression – may occur at young age, as in the reported adolescent patients of African descent.
P62  **A clinical classification system for Paravenous Pigmentary Chorioretinal Atrophy (PPCRA) in Childhood**

Göran Darius Hildebrand¹, Dorothy Thompson², Isabelle Russell-Eggitt², David Taylor²

¹ King Edward VII Hospital, Windsor, UK, ² Great Ormond Street Hospital, London, UK

See: Rapid Fire RF29.

P63  **Choroidal neovascularization secondary to Autosomal Recessive Bestrophinopathy in a 9-year-old girl treated by intravitreal ranibizumab injection**

Marta Pawlak¹, Anna Gotz-Wieckowska¹, Joanna Siwiec-Proscinska¹

¹ Department of Ophthalmology University of Medical Sciences Poznan, Poland

See: Rapid Fire RF30.

P64  **Identical Twins Exposed! – Bestrophinopathy, Autosomal Recessive?**

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See: Rapid Fire RF31.

P65  **Laser flare photometry in healthy preschool children**

Judit Kőrtvélyes¹, Maria Bausz¹, Bela Csakany¹, Szabolcs Benedek¹, Janos Nemeth¹

¹ Semmelweis University, Department of Ophthalmology, Budapest, Hungary

See: Rapid Fire RF33.

P66  **Megalopapilla in children: definition by OCT**

Elena Jarrín Hernández¹, Laura Cabrejas Martínez², Luciano Bravo Ljubetic³, Soraya Mediero Clemente³, Susana Noval Martínez³

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See: Rapid Fire RF32.
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# Topic and location of previous annual meetings

**European Paediatric Ophthalmological Society (EPOS) 2001 – 2013**

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<thead>
<tr>
<th>Year</th>
<th>Location</th>
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<td>2012</td>
<td>Uppsala, Sweden</td>
<td>Developments in Paediatric Ophthalmology</td>
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<tr>
<td>2011</td>
<td>Thessaloniki, Greece</td>
<td>Visual Impairment in Childhood</td>
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<td>2010</td>
<td>Bad Nauheim, Germany</td>
<td>New Challenges in Paediatric Ophthalmology</td>
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<td>Paris, France</td>
<td>Perinatal Ophthalmology</td>
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<td>2008</td>
<td>Leuven, Belgium</td>
<td>The Eye in Systemic Disease</td>
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<td>2007</td>
<td>Portoroz, Slovenia</td>
<td>Paediatric Electrophysiology and Psychophysics</td>
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<td>2006</td>
<td>Vilamoura, Portugal</td>
<td>Pediatric Neuro-ophthalmology</td>
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<td>2005</td>
<td>Warszaw, Poland</td>
<td>Advances in the surgical treatment of pediatric eye diseases</td>
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<td>2004</td>
<td>Manchester, UK</td>
<td>Developmental genes and the eye</td>
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<td>2003</td>
<td>Regensburg, Germany</td>
<td>Gene Therapy and Other Modern Therapeutic Approaches in Paediatric Retinal Degenerations</td>
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<td>2002</td>
<td>Figuera davFo, Portugal</td>
<td>Dysmorphology of the Eye and Orbit</td>
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<td>2001</td>
<td>Regensburg, Germany</td>
<td>Trends in Paediatric Ophthalmology</td>
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**European Paediatric Ophthalmology Group (EPOG) 1973 – 2000**

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<td>2000</td>
<td>Cambridge, UK</td>
<td>Retinal dystrophies</td>
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<td>Strasbourg, France</td>
<td>Multisystem disease and the eye</td>
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<td>Metabolic diseases of the eye</td>
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<td>Sandbjerg, Denmark</td>
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<td>Cataract in childhood</td>
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<td>Copenhagen, Denmark</td>
<td>Visually handicapped children the ophthalmologist's responsibility</td>
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<td>1973</td>
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<td>Retinitis pigmentosa</td>
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Leiden, the Netherlands, October 11-12, 2013
Scientific Committee

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and:

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Dr. Liesbeth Prick

This meeting would not have been possible without the valuable support of:

Caroline van Battum and Jacob Muller from Boerhaave Congress Organisation
Annelies Donkersloot, abstract book and secretarial support
Corrie Hindaal, organization of reception and dinner
Heiko Klarl, the webmaster
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