PATIENT INVOLVEMENT IN THE DEVELOPMENT OF SURGICAL CONSENSUS STATEMENTS FOR ESOPHAGEAL ATRESIA (EA) DISEASE ERNICA

July 4, 2019
Anke Widenmann-Grolig, Treasurer of EAT

- Thank you *Dr. Carmen Dingemann*, MHH for contributing her presentation which was a huge workload.
- Thank you PhD *Simon Eaton* for all the scientific and technical work
- Thank you Prof. Benno Ure for providing this to the patient community

EAT
– The Federation of Esophageal Atresia and Tracheo-Esophageal Fistula Support Groups –
www.we-are-eat.org
EAT - Who we are...

- A federation whose members are national patient support groups for EA
- Founded in 2011
- Legally registered in Stuttgart as an ‘e.V.’
- International but mainly European
- Seven Board members (elected by the members)
- Has its own Medical Advisory Board (INoEA - founded by Frédéric Gottrand)

EAT and INoEA could be described as an international ‘family’!

First Esophageal Atresia Award (EWA) in 2019 for Prof. Frédéric Gottrand, Lille
5th World Congress of Esophageal Atresia in Rome, Italy
Hospitals in ERNICA

There are 20 HCPs (hospitals) in the ERNICA network from 10 countries:

- Belgium
- Denmark
- Finland
- France
- Germany
- Italy
- Netherlands
- Norway
- Sweden
- UK

ERNICA is coordinated by Prof. Dr. René Wijnen (Head of Paediatric Surgery) at Erasmus MC, Rotterdam, The Netherlands
European Reference Networks on Inherited and Congenital Anomalies

- Esophageal diseases
- Intestinal diseases
- Intestinal failure
- Gastroenterological diseases
- Malformations of the diaphragm and abdominal wall
ePAGs in ERNICA

- Esophageal diseases  Graham, JoAnne, Anke
- Intestinal diseases  Annette Lemli & Nicole Schwarzer (SoMA)
  cross link to ernUrogen
- Intestinal failure  Nadine & Sylvia (do not continue)
- Gastroenterological diseases  NEW (no ePAGs)
- Malformations of the diaphragm and abdominal wall  Beverley Power, Fanny Cauvet
European Reference Networks on Inherited and Congenital Anomalies (ERNICA)

Network Activities

- Standards of Care
- Research
- Training
- Registry

ePAG involvement in every working group
Background

ERNICA

April 2017, Nov 2017, April 2018
ERNICA meetings in Rotterdam, Helsinki, Stockholm
Always with participation of patient representatives

- Little evidence on current diagnostic and therapeutic concepts
- Generally accepted guidelines and algorithms lacking

Decision of Workstream
Congenital Malformations and Diseases of the Esophagus

Consensus Conference
on the Perioperative and Surgical Management of Patient with Esophageal Atresia with Tracheoesophageal Fistula
Follow-up was the additional topic of the ePAGs
Additional Values by Patient Representatives

- Including Follow-Up and Transition already in the first consensus conference
- Bridging the existing gap between consensus statements and patient needs by implementing a patient journey - ePAGs have the lead.
- No double work as Dr. Carmen Dingemann is working in both groups
- Patient journey can use the results of the consensus conference for the internal work before publication
Background

**ERNICA Consensus Conference**

Berlin, 25th and 26th October 2018

Anke Widenmann-Grolig  
KEKS Germany & EAT

Graham Slater  
TOFS, UK & EAT

JoAnne Fruithof  
VOKS NL & EAT
Participants (n = 19)

Members of ERNICA Workstream
Congenital Malformations and Diseases of the Esophagus

- Pediatric surgeon: 14
- Pediatric gastroenterologist: 1
- Methodologist: 1
- Patient support group represent.: 3

9 countries
**Methods**

**ERNICA Consensus Conference**

**Preparation of the Conference**

1. Literature search -> CD, SE, BM
2. Item generation
   - Including Patient representatives -> Workstream members (Stockholm) -> CD, SE, BM
3. Item prioritization
   - Including Patient representatives -> Conference participants
     - 5-point Likert scale online survey
4. Final list containing relevant domains
   - diagnostics, preoperative, operative and postoperative management, follow-up*, varia

*Follow-up* based on ESPGHAN-NASPGHAN Guidelines

## Part I

### Section I
- **Domain**: Diagnostics

### Section II
- **Domain**: Preoperative Management

### Section III
- **Domain**: Operative Management 1

### Section IV
- **Domain**: Operative Management 2

### Section V
- **Domain**: Postoperative Management

## Part II

### Section VI
- **Domain**: Follow-up 1

### Section VII
- **Domain**: Follow-up 2

### Section VIII
- **Domain**: Varia
Methods

**ERNICA Consensus Conference**

1. Discussion based on the highest available level of evidence of current literature

2. Anonymous voting / internet-based system / 1-9 scale

   - 1 Strongly Disagree
   - 2
   - 3
   - 4
   - 5
   - 6
   - 7
   - 8
   - 9 Fully Agree

3. Definition of consensus

   > 75% of those voting scoring 6, 7, 8, 9
Example of Voting via [VoxVote](https://www.voxvote.com/)

1. Should a nasogastric tube be routinely inserted as a diagnostic procedure in cases with suspected EA?

   - 9 - Fully Agree: 15.8%
   - 8: 15.8%
   - 7: 10.5%
   - 6: 5.3%
   - 5: 5.3%
   - 4: 5.3%
   - 3: 10.5%
   - 2: 26.3%
   - 1 - Strongly Disagree: 5.3%

19 users voted
In accordance with the Oxford CEBM Levels of Evidence as published in 2009

Consensus Conference Part I

Results

116 relevant publications
5 (4.3%) Level-1-evidence studies

Percentage of included articles (n = 116)
Results

Consensus Conference Part I

Preoperative Management*

<table>
<thead>
<tr>
<th>Item</th>
<th>Description</th>
<th>Consensus</th>
<th>%</th>
<th>Votes</th>
<th>Median [range]</th>
</tr>
</thead>
<tbody>
<tr>
<td>7</td>
<td>A replogle tube should be routinely placed into the upper esophageal pouch to allow continuous low pressure suction.</td>
<td>+</td>
<td>100</td>
<td>15/15</td>
<td>9 [6-9]</td>
</tr>
<tr>
<td>8</td>
<td>Preoperative antibiotic prophylaxis should be routinely administered as soon as the diagnosis is established.</td>
<td>-</td>
<td>13.3</td>
<td>2/15</td>
<td>2 [1-9]</td>
</tr>
<tr>
<td>9</td>
<td>Spontaneous breathing should routinely be favoured.</td>
<td>+</td>
<td>100</td>
<td>15/15</td>
<td>9 [9-9]</td>
</tr>
<tr>
<td>10</td>
<td>If assisted ventilation is required, intubation should be preferred to non-invasive ventilation.</td>
<td>+</td>
<td>100</td>
<td>15/15</td>
<td>9 [8-9]</td>
</tr>
<tr>
<td>11</td>
<td>Tracheobronchoscopy under spontaneous breathing should be performed preoperatively to evaluate tracheomalacia.</td>
<td>-</td>
<td>53.3</td>
<td>8/15</td>
<td>6 [2-9]</td>
</tr>
<tr>
<td>12</td>
<td>A central venous line should be routinely placed preoperatively.</td>
<td>-</td>
<td>14.3</td>
<td>2/14</td>
<td>2 [1-7]</td>
</tr>
<tr>
<td>13</td>
<td>An arterial line should be routinely placed preoperatively.</td>
<td>-</td>
<td>7.1</td>
<td>1/14</td>
<td>1 [1-8]</td>
</tr>
<tr>
<td>14</td>
<td>During preoperative counselling parents should be routinely informed about different surgical options such as open and thoracoscopic repair.</td>
<td>+</td>
<td>94.4</td>
<td>17/18</td>
<td>9 [2-9]</td>
</tr>
</tbody>
</table>

*before the patient is transferred to operation theatre
# Consensus Conference Part I

## Operative Management

<table>
<thead>
<tr>
<th>OPERATIVE MANAGEMENT</th>
<th>Consensus</th>
<th>%</th>
<th>Votes</th>
<th>Median [range]</th>
</tr>
</thead>
<tbody>
<tr>
<td>15. A stable neonate with EA should preferably be operated during working hours</td>
<td>+</td>
<td>94.4</td>
<td>17/18</td>
<td>9 [3-9]</td>
</tr>
<tr>
<td>during the week.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>16. Antibiotics should be routinely administered perioperatively.</td>
<td>+</td>
<td>100</td>
<td>14/14</td>
<td>9 [8-9]</td>
</tr>
<tr>
<td>17. A central venous line should be placed before the operation.</td>
<td>+</td>
<td>93.3</td>
<td>14/15</td>
<td>9 [1-9]</td>
</tr>
<tr>
<td>18. An arterial line should be placed before the operation.</td>
<td>+</td>
<td>76.8</td>
<td>11/14</td>
<td>8 [1-9]</td>
</tr>
<tr>
<td>19. Tracheoscopy should be routinely performed before the operation to evaluate</td>
<td>+</td>
<td>94.1</td>
<td>16/17</td>
<td>9 [2-9]</td>
</tr>
<tr>
<td>the fistula(s) and other tracheolaryngeal pathology.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>20. Horizontal or vertical or U-shaped (Bianchi) approaches (skin incision) are</td>
<td>+</td>
<td>100</td>
<td>15/15</td>
<td>9 [8-9]</td>
</tr>
<tr>
<td>viable approaches for conventional thoracotomy.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>21. Muscle-sparing approach is the recommended approach for conventional thoracotomy.</td>
<td>+</td>
<td>100</td>
<td>15/15</td>
<td>9 [8-9]</td>
</tr>
<tr>
<td>22. Entry through the 4th intercostal space is the recommended approach for</td>
<td>+</td>
<td>100</td>
<td>14/14</td>
<td>9 [7-9]</td>
</tr>
<tr>
<td>conventional thoracotomy.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>23. The extrapleural approach is the preferred approach for thoracotomy.</td>
<td>+</td>
<td>92.9</td>
<td>13/14</td>
<td>9 [5-9]</td>
</tr>
<tr>
<td>24. In cases with suspected right descending aorta, a right-sided thoracic approach</td>
<td>+</td>
<td>76.9</td>
<td>10/13</td>
<td>8 [1-9]</td>
</tr>
<tr>
<td>is the first option.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>25. The azygos vein should be preserved whenever possible.</td>
<td>-</td>
<td>71.4</td>
<td>10/14</td>
<td>6.5 [2-9]</td>
</tr>
<tr>
<td>26. The thoracoscopic approach is a viable option.</td>
<td>+</td>
<td>87.5</td>
<td>14/16</td>
<td>9 [5-9]</td>
</tr>
<tr>
<td>27. The thoracoscopic approach should be only performed where suitable expertise is</td>
<td>+</td>
<td>100</td>
<td>17/17</td>
<td>9 [6-9]</td>
</tr>
<tr>
<td>available.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>28. The thoracoscopic approach offers the advantage of magnification compared to the</td>
<td>+</td>
<td>92.3</td>
<td>13/14</td>
<td>9 [5-9]</td>
</tr>
<tr>
<td>conventional approach.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
**Consensus Conference Part I**

### Postoperative Management

<table>
<thead>
<tr>
<th>Item</th>
<th>Description</th>
<th>Consensus</th>
<th>%</th>
<th>Votes</th>
<th>Median [range]</th>
</tr>
</thead>
<tbody>
<tr>
<td>43</td>
<td>Postoperative ventilation and relaxation should not be routine and reserved for selected patients such as those with tension anastomosis.</td>
<td>+</td>
<td>100</td>
<td>14/14</td>
<td>9 [6-9]</td>
</tr>
<tr>
<td>44</td>
<td>Routine postoperative antibiotic treatment for longer than 24 hours should be recommended.</td>
<td>-</td>
<td>13.3</td>
<td>2/15</td>
<td>2 [1-9]</td>
</tr>
<tr>
<td>45</td>
<td>A postoperative contrast study of the esophagus should be routinely performed before the initiation of oral feeding.</td>
<td>-</td>
<td>20</td>
<td>3/15</td>
<td>1 [1-7]</td>
</tr>
<tr>
<td>46</td>
<td>Feeding via the transanastomotic tube may be routinely initiated before 24 hours postoperatively.</td>
<td>+</td>
<td>100</td>
<td>15/15</td>
<td>9 [7-9]</td>
</tr>
<tr>
<td>47</td>
<td>Oral feeding may be routinely initiated after 24 hours postoperatively.</td>
<td>+</td>
<td>100</td>
<td>15/15</td>
<td>9 [6-9]</td>
</tr>
<tr>
<td>48</td>
<td>An anastomotic leakage should be routinely managed with a chest drain.</td>
<td>+</td>
<td>92.9</td>
<td>15/15</td>
<td>8 [6-9]</td>
</tr>
<tr>
<td>49</td>
<td>An anastomotic leakage within the first 4 postoperative days may be considered for surgical revision.</td>
<td>-</td>
<td>71.4</td>
<td>10/14</td>
<td>8 [2-9]</td>
</tr>
<tr>
<td>50</td>
<td>A contrast study, tracheoscopy and esophagoscopy are necessary to exclude a re-fistula or missed upper pouch fistula, if suspected.</td>
<td>+</td>
<td>93.8</td>
<td>15/16</td>
<td>9 [3-9]</td>
</tr>
<tr>
<td>51</td>
<td>A re-fistula may be initially managed by either endoscopic or surgical approach.</td>
<td>+</td>
<td>100</td>
<td>14/14</td>
<td>9 [6-9]</td>
</tr>
<tr>
<td>52</td>
<td>A clinical checklist should be made available including items which should be performed before first discharge (i.e. abdominal and renal ultrasound, resuscitation training for parents/caregivers).</td>
<td>+</td>
<td>100</td>
<td>18/18</td>
<td>9 [9-9]</td>
</tr>
</tbody>
</table>
Consensus Conference Part II

Part II – Follow-up: More topics patients were competent e.g. 1. - life-long follow-up was key for us

Results will be published by the end of 2019
ERNICA Consensus Conference on the Perioperative and Surgical Management of Patients with Esophageal Atresia with Tracheoesophageal Fistula

Dingemann C¹, Eaton S², Aksnes G³, Bagolan P⁴, Cross K⁵, De Coppi P⁵, Fruithof J⁶, Gamba P⁷, Husby S⁸, Koivusalo A⁹, Rasmussen L¹⁰, Sfeir R¹¹, Slater G¹², Svensson JF¹³, Van der Zee D¹⁴, Wessel L¹⁵, Widenmann-Grolig A¹⁶, Wijnen R¹⁷, Ure B¹

¹Department of Pediatric Surgery, Hannover Medical School, Hannover, Germany
²Paediatric Surgery & Metabolic Biochemistry, UCL Great Ormond Street Institute of Child Health, London, UK
³Department of Pediatric Surgery, Oslo University Hospital, Oslo, Norway
⁴Neonatal Surgery Unit, Bambino Gesù Children’s Hospital-Research Institute Rome, Rome, Italy
⁵Specialist Neonatal and Paediatric Surgery, Great Ormond Street Hospital for Children, London, UK
⁶Esophageal Atresia and Tracheo-Esophageal Fistula Support Group VOKS, Lichtenvoorde, Netherlands
⁷Department of Pediatric Surgery, University of Padua, Padua, Italy
⁸Department of Pediatric Gastroenterology, Odense University Hospital, Odense, Denmark
⁹Department of Pediatric Surgery, University of Helsinki, Helsinki, Finland
¹⁰Department of Pediatric Surgery, Odense University Hospital, Odense, Denmark
¹¹Department of Pediatric Surgery, Centre Hospitalier Regional Universitaire de Lille, Lille, France
¹²Esophageal Atresia and Tracheo-Esophageal Fistula Support Group TOFS, Nottingham, United Kingdom
¹³Department of Pediatric Surgery, Karolinska University Hospital, Stockholm, Sweden
¹⁴Department of Pediatric Surgery, Utrecht University Medical Center, Utrecht, Netherlands
¹⁵Department of Pediatric Surgery, University of Mannheim, Medical Faculty of Heidelberg, Mannheim, Germany
¹⁶Esophageal Atresia and Tracheo-Esophageal Fistula Support Group KEKS, Stuttgart, Germany
¹⁷Department of Pediatric Surgery, Erasmus MC Rotterdam, Rotterdam, Netherlands

All authors are representatives of the European Reference Network for Rare Inherited Congenital Anomalies (ERNICA).
Success factors

- Support of Coordinator (René Wijnen and work-stream clinical leads (Frédéric Gottrand & Benno Ure)
- Positive relationships at national level e.g. Carmen Dingemann
- Soft skills of ePAGs and financial support by national organizations
- Very good communication and relationship amongst the ePAG
- The ePAG advocates were involved in the whole process
- The ePAGs disseminate the results into the national groups to inform the clinicians in all countries even when there are no members of ERNICA.
- Fast dissemination: EUPSA Congress in June 2019 and during the 5th World Congress of Esophageal Atresia at the end of June 2019 which helped to disseminate the results quicker.
There is no substitute for a close and trusting relationship between clinicians and ePAG advocates, but building this trust takes time (10 years for EAT).

The question “how do ePAG advocates make sure that they bring a relevant/evident opinion for all patients of this special group”? is not so easy to be trained. Experience in the patient work is compulsory or a good back office team.

Coordinator support is crucial in ‘setting the tone’ for the ERN.
Lessons learned - 2

- The use of a recognized methodology also helps to generate consensus statements which have the potential to be disseminated as ‘best practice’ when there is a lack of scientific ‘evidence’ as would be normally demanded.

- We learned how difficult and complex it is to develop an evident consensus statement (first level of evidence).
We thank all the clinicians of the consensus conference and their engagement. Special thanks to Carmen and Simon who had the major work load. Many thanks to René as our Coordinator and Frédéric and Benno for their openminded leading of the workstream.

ePAGs Anke, Graham, JoAnne