

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 **E**sophageal **A**tresia and
Tracheo-**E**sophageal **F**istula 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



ERNICA

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



ERNICA

European Reference Networks on Inherited and Congenital Anomalies

Network Activities

ePAG involvement in every working group

Standards
of Care

Research

Training

Registry

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



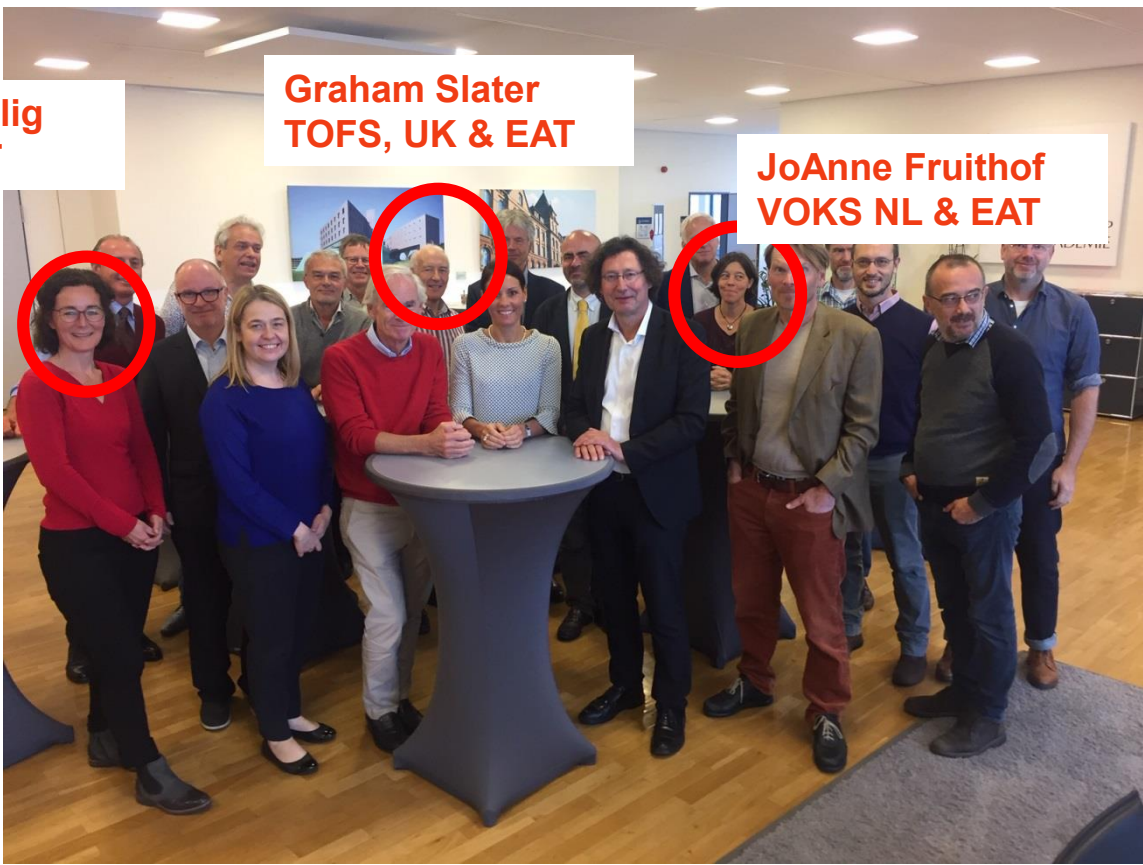
ERNICA Consensus Conference

Berlin, 25th and 26th October 2018

Anke Widenmann-Grolig
KEKS Germany & EAT

Graham Slater
TOFS, UK & EAT

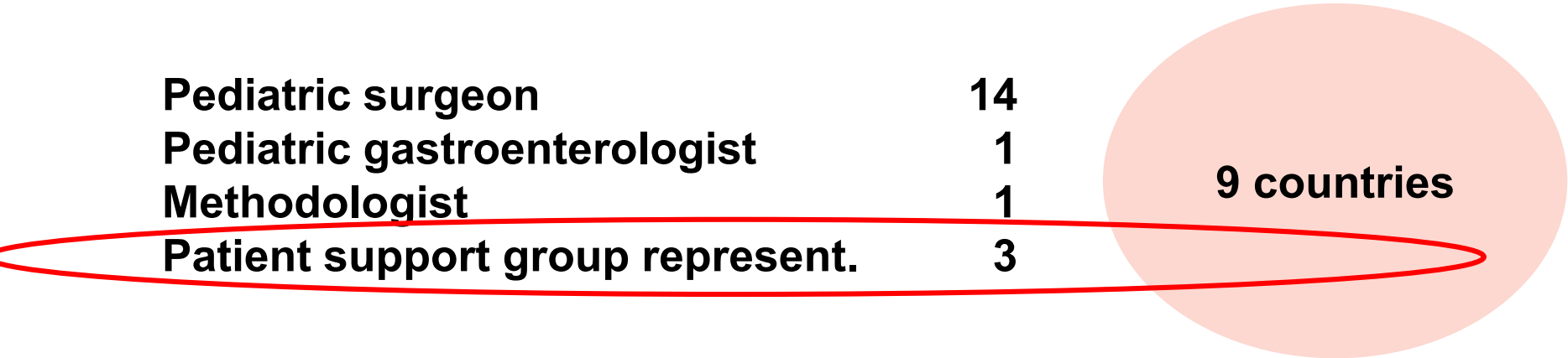
JoAnne Fruithof
VOKS NL & EAT



ERNICA Consensus Conference

Participants (n = 19)

Members of ERNICA Workstream
Congenital Malformations and Diseases of the Esophagus



ERNICA Consensus Conference

Preparation of the Conference

1

Literature search -> CD, SE, BM

2

Item generation -> Workstream members (Stockholm)
Including Patient representatives -> CD, SE, BM

3

Item prioritization -> Conference participants
Including Patient representatives 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
Krishnan U et al. J Pediatr Gastroenterol Nutr. 2016*

ERNICA Consensus Conference

Preparation of the Conference

Section	Domain
I	Diagnostics
II	Preoperative Management
III	Operative Management 1
IV	Operative Management 2
V	Postoperative Management
VI	Follow-up 1
VII	Follow-up 2
VIII	Varia

Part I

Part II

ERNICA Consensus Conference

Voting

Including Patient
representatives in Berlin

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

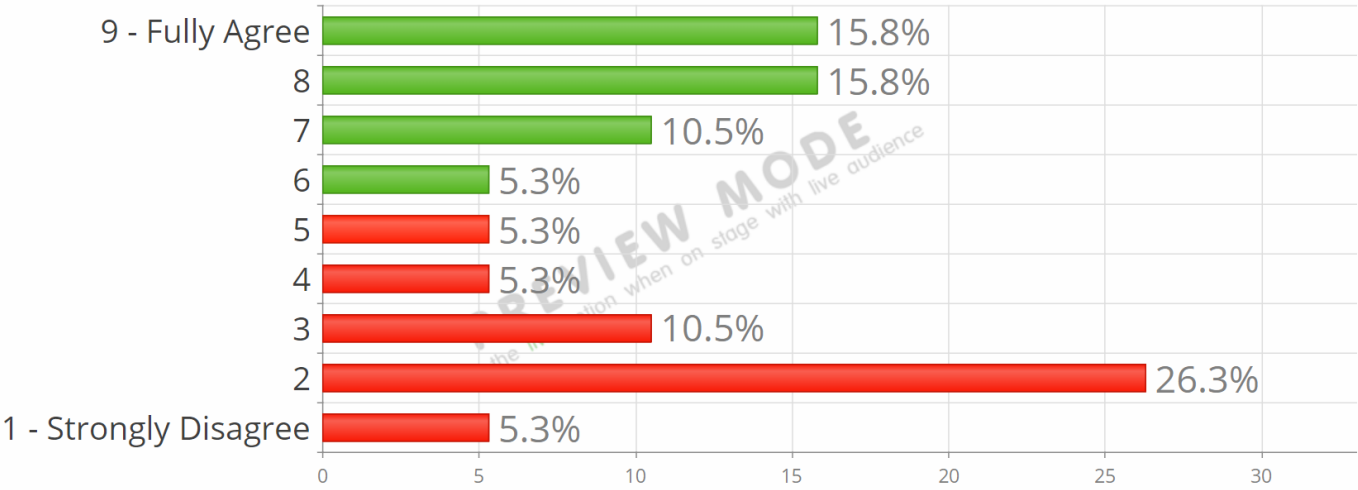
Definition of consensus

> 75% of those voting scoring 6, 7, 8, 9

ERNICA Consensus Conference

Example of Voting via VOXvote

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

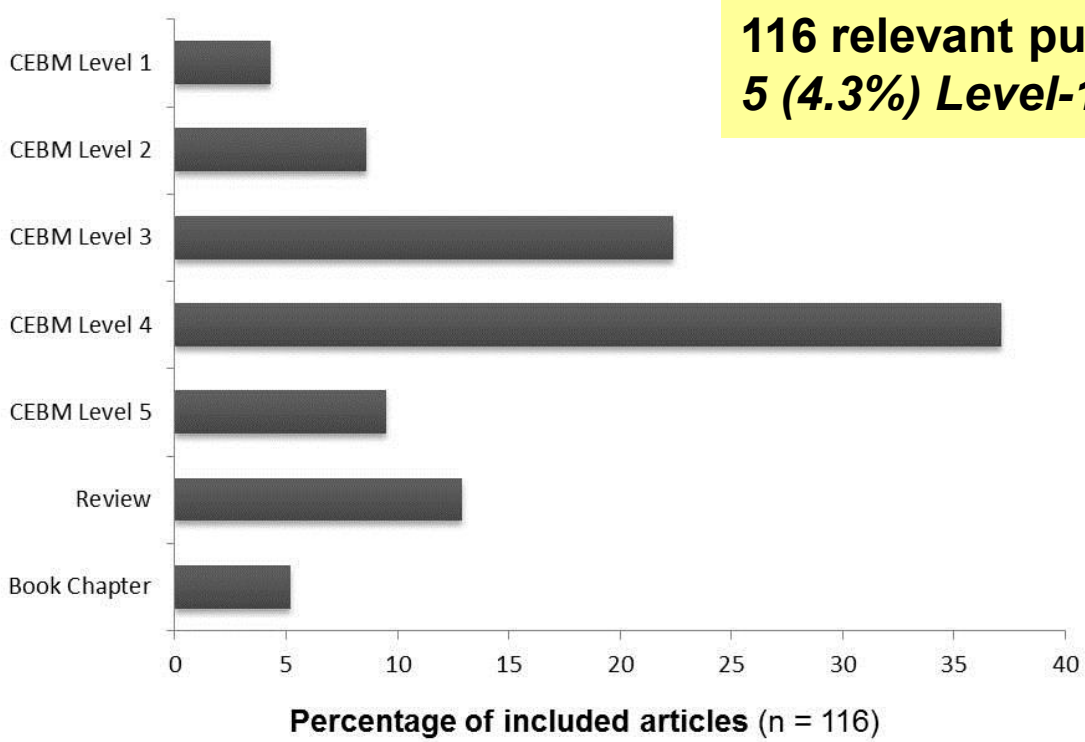


19
users
voted

<https://www.voxvote.com/>

Consensus Conference **Part I**

Literature



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

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

Consensus Conference **Part I**

Preoperative Management*

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

**before the patient is transferred to operation theatre*

Consensus Conference **Part I**

Operative Management

	OPERATIVE MANAGEMENT	Consensus	%	Votes	Median [range]
15	A stable neonate with EA should preferably be operated during working hours during the week.	+	94.4	17/18	9 [3-9]
16	Antibiotics should be routinely administered perioperatively.	+	100	14/14	9 [8-9]
17	A central venous line should be placed before the operation.	+	93.3	14/15	9 [1-9]
18	An arterial line should be placed before the operation.	+	78.8	11/14	8 [1-9]
19	Tracheoscopy should be routinely performed before the operation to evaluate the fistula(s) and other tracheolaryngeal pathology.	+	94.1	16/17	9 [2-9]
20	Horizontal or vertical or U-shaped (Bianchi) approaches (skin incision) are viable approaches for conventional thoracotomy.	+	100	15/15	9 [7-9]
21	Muscle-sparing approach is the recommended approach for conventional thoracotomy.	+	100	15/15	9 [8-9]
22	Entry through the 4 th intercostal space is the recommended approach for conventional thoracotomy.	+	100	14/14	9 [7-9]
23	The extrapleural approach is the preferred approach for thoracotomy.	+	92.9	13/14	9 [5-9]
24	In cases with suspected right descending aorta, a right-sided thoracic approach is the first option.	+	76.9	10/13	8 [1-9]
25	The azygos vein should be preserved whenever possible.	-	71.4	10/14	6.5 [2-9]
26	The thoracoscopic approach is a viable option.	+	87.5	14/16	9 [5-9]
27	The thoracoscopic approach should be only performed where suitable expertise is available.	+	100	17/17	9 [6-9]
28	The thoracoscopic approach offers the advantage of magnification compared to the conventional approach.	+	92.9	13/14	9 [5-9]

Consensus Conference **Part I**

Postoperative Management

	POSTOPERATIVE MANAGEMENT	Consensus	%	Votes	Median [range]
43	Postoperative ventilation and relaxation should not be routine and reserved for selected patients such as those with tension anastomosis.	+	100	14/14	9 [6-9]
44	Routine postoperative antibiotic treatment for longer than 24 hours should be recommended.	-	13.3	2/15	2 [1-9]
45	A postoperative contrast study of the esophagus should be routinely performed before the initiation of oral feeding.	-	20	3/15	1 [1-7]
46	Feeding via the transanastomotic tube may be routinely initiated at 24 hours postoperatively.	+	100	15/15	9 [7-9]
47	Oral feeding may be routinely initiated after 24 hours postoperatively.	+	100	15/15	9 [6-9]
48	An anastomotic leakage should be routinely managed with a chest drain.	+	92.9	13/14	8 [2-9]
49	An anastomotic leakage within the first 4 postoperative days may be considered for surgical revision.	-	71.4	10/14	8 [1-9]
50	A contrast study, tracheoscopy and esophagoscopy are necessary to exclude a re-fistula, or missed upper pouch fistula, if suspected.	+	93.8	15/16	9 [3-9]
51	A re-fistula may be initially managed by either endoscopic or surgical approach.	+	100	14/14	9 [6-9]
52	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).	+	100	18/18	9 [9-9]

Consensus Conference **Part II**

	Follow-up	Consensus	%	Votes	Median [range]
1	There should be a structured schedule for life-long follow-up	+	100	18/18	9 [8-9]
<div><div>Part II – Follow-up: More topics patients were competent</div><div>e.g. 1. - life-long follow-up was key for us</div><div>Results will be published by the end of 2019</div></div>				18/18	9 [8-9]
				16/18	9 [2-9]
				16/16	8 [6-9]
				12/18	7 [1-9]
				17/18	9 [4-9]
				17/18	8.5 [3-9]
				15/15	9 [6-9]
				17/18	9 [4-9]
				15/18	8 [1-9]
				4/15	3 [1-9]
				6/13	5 [3-9]
				14/14	8 [6-9]
				15/16	9 [5-9]

delines available

ERNICA Consensus Conference

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

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¹⁶Esophageal Atresia and Tracheo-Esophageal Fistula Support Group KEKS, Stuttgart, Germany

¹⁷Department of Pediatric Surgery, Erasmus MC Rotterdam, Rotterdam, Netherlands

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.



Lessons learned -1

- 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