

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

June 2019

Activity	<i>Patient involvement in the development of surgical consensus statements for esophageal atresia (EA) disease</i>
Area	<i>Clinical practice guidelines and healthcare, quality and safety</i>
Duration	<i>April 2018 – Q1 2020</i>
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## 1. SUMMARY

**i** A set of 5 consensus statements for **Diagnostics, Preoperative, Operative, Postoperative Management and Follow-up of Esophageal Atresia (EA<sup>1</sup>) and Tracheoesophageal fistula (TEF)** has been developed by ERNICA ERN to provide a more consistently higher standard of treatment and care for patients with EA and TEF.

For each of the 5 consensus domains, the clinicians together with the patients went through the following process: (i) item generation, (ii) item prioritization (iii) formulation of a final list of items (iv) evidence check for each of the domains. ePAG advocates were included in the discussion with the clinicians at a very early stage of the project development.

A 2-day consensus conference was organized in Berlin in October 2018 to work on the first consensus statements in the work stream *Diseases of the Esophagus*. After an extensive literature search and preparation process that took approximately 9 months, relevant aspects with regard to diagnostic, pre-, intra-, and postoperative management as well as regarding follow-up of patients with EA/TEF were discussed, and

<sup>1</sup> Esophageal Atresia (EA)/Tracheoesophageal fistula is a congenital malformation resulting from abnormal development before birth of the tube that carries food from the mouth to the stomach. It affects approx. 1 in 3500 births. Both diseases fall under the scope of the ERNICA work-stream 'Diseases of the Esophagus'.

consensus was reached on 81 items after an anonymous online voting process<sup>2</sup>. Participants to this meeting included 15 clinicians, 1 healthcare researcher and 3 ePAG advocates from 9 European countries, all involved in ERNICA.

A structured methodology and anonymous online voting was used throughout the meeting to prioritise the items and reach consensus (consensus was defined when  $\geq 75\%$  of the votes scored 6-9 in a 1-9 scale). ePAG advocates participated to the discussion and voted on several items (see Annex I).

The first 52 consensus statements have been published in the *'European Journal of Pediatric Surgery'* (except the 29 items for the Follow-up domain which will be published by the end of 2019). The ePAG Advocates involved in this work are acknowledge as co-authors of this paper: "ERNICA Consensus Conference On The Management Of Patients With Esophageal Atresia And Tracheoesophageal Fistula: Diagnostics, Preoperative, Operative And Postoperative Management": <http://discovery.ucl.ac.uk/10076151/>

## 2. INITIAL SITUATION

**i** There is a wide variation in outcomes for EA patients that could potentially be explained by a variety of factors, including the surgical approach, the surgeon, the treatment, the country, etc. However, a systematic comparison of results is not possible if we do not have comparable surgical approaches. The lack of a common evidence-based guidance for surgical management of newborns with EA prevents benchmarking and leads to disparate health outcomes for this patient community.

During its kick-off meeting in 2017, ERNICA clinicians already identified the need to develop a consensus statements for EA, initially focusing on the surgical aspects. We, as patient representatives, were involved in the development of the consensus statements since these initial discussions and we emphasized the need of including also other aspects such as the diagnostics, the postoperative management and in particular we convinced them to introduce the follow-up.

The ePAG advocates involved in this project are from France, Germany and The Netherlands, and are all Board members of EAT (the International Federation of EA patient support groups). These ePAG advocates already had a good working relationships with many of the 15 clinicians involved in this exercise which has proved very beneficial to the progress made. They have worked together for over 30 years at national and international level in a very trustful surrounding. EAT was already collaborating with a loose network of doctors (INoEA = International Network of Esophageal Atresia) since 2010. In addition, most of the European Steering Committee members, the medical board of EAT, are part of the ERNICA network. This international collaboration of nearly one decade has been key to progress fast in this project and instrumental to engage patient representatives as partners.

## 3. CONTRIBUTION TO PATIENT ENGAGEMENT AND/OR IMPROVEMENT OF CARE

**i** This project is a prime example of good collaborative patient involvement in the ERNICA network.

**ePAG advocates have been actively involved throughout the whole project, not only at the voting phase:**

- We contributed to identify the publications for the literature review.
- We co-led the process on the Follow-up domain: we convinced the clinicians to include a 5<sup>th</sup>

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<sup>2</sup> ERNICA Newsletter, January 2019

consensus statement on follow-up and we consulted the patient support groups who had reliable publications of their national medical board about the aftercare. In total there were only 4 evident publications for all items (see Annex II), which show the need for the consensus guideline as the first level of evidence.

- We participated in the definition of the deliverables on guidelines as top priority for ERNICA in year 2.
- We contributed to the definition of the **Delphi voting process**<sup>3</sup> to set up the priorities per item: we suggested additional wording (*recommend* instead of *inform about* patient support group) and we added the checklist before the first release from hospital.
- We were considered as equal partners with the same rights during the voting process.

**The consensus addresses several of our needs as patient groups:**

- Better diagnosis and surgery when bronchoscopy *before* surgery
- Better preparation for parents for the coming challenges after release (checklist)
- Chance for a second opinion before initial surgery (first shot is the most important one)

**This project has great potential to improve the treatment of EA patients** (newborns with only one single chance for a good primary repair) prior to, during and in the immediate post-operative phase. The commitment on a lifelong follow-up and transition which will be published in the second part, will help us to implement adequate treatment for the time after surgery.

**These consensus statements are also complementary to the work of the 'ERNICA 'Patient Journey' project**, as we can use many of the consensus items in our patient journey. It is a matter of patient centered language to reach more parents and patients.

Broad uptake of the consensus across European countries will lead to reductions in morbidity and contribute to standardize and improve care for these patients. The challenge now, as is generally the case, will be to get these consensus statements and improved care standards which will emerge from this project adopted and implemented across all countries.

Right after the publication of the first consensus statement in June 2019, **the ePAG advocates could convince the clinicians to prepare a second consensus statement for the management of long-gap esophageal atresia**. This subtype of disease represents only 10% of all cases, but the management of these patients is usually very difficult and even more complex. As those patients can be easily transferred into another hospital we hope for clear recommendation for a centralized surgery and a de-centralized aftercare with the support from the surgeons of the expert center.

## 4. SUCCESS FACTORS

- i** • The support of the ERNICA Coordinator and work-stream clinical leads, with a clear vision of successful clinician-patient partnership, has helped to be seen as equal partners in the development of this project.
- The pre-existing positive relationships at national level with relevant clinicians participating in this project was essential.
- Soft skills of the patients and financial support provided by the national organization groups were also

<sup>3</sup> The Delphi method is a structured communication technique or method, originally developed as a systematic, interactive forecasting method which relies on a panel of experts. Delphi is based on the principle that forecasts (or decisions) from a structured group of individuals are more accurate than those from unstructured groups. The experts answer questionnaires in two or more rounds. After each round, a facilitator or change agent provides an anonymised summary of the experts' forecasts from the previous round as well as the reasons they provided for their judgments.

part of the success. There was a very good communication and relationship amongst the ePAG advocates involved who also brought into the project good management and operational skills.

- The ePAG advocates were involved in the whole process and promote a more patient centered approach: we were on the mailing list as every clinician. We had three voices out of 18. We had the possibility to express our opinion (and say that “we have no expertise on this specific matter”). Some clinicians used this possibility as well to acknowledge their own limitations. We discussed the questions and the priorities as a group before we made our votes, so we made concrete comments and we brought our patients’ perspective (see example in Annex I).
- The ePAG advocates participated in the dissemination of the results and will support the process of implementation. We disseminate the results into our national groups to inform the clinicians in all countries even when there are no members of ERNICA.
- The results were presented at the EUPS Congress in June 2019 and during the 5<sup>th</sup> World Congress of Esophageal Atresia at the end of June 2019 which helped to disseminate the results quicker.

## 5. LESSONS LEARNED

- i** • There is no substitute for a close and trusting relationship between clinicians and ePAG advocates if we are to make a full contribution to the work of an ERN, but building this trust takes time.
- It is important, that the ePAG advocates are either well informed in their national network about the problems and needs of the patients or are keen on exchanging with others. 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. This is a task for the European Federation for a specific disease. Without our broad knowledge within our national group and the very close collaboration amongst us, this would not have been possible. EAT already conducted a Patient Reported Outcomes Measures (PROM) survey in 5 languages and received 1.000 patient answers in 2014 – so we had a very good overlook about the questions with the highest priority from patient side.
- Coordinator support is crucial in ‘setting the tone’ for the ERN as regards patient involvement. A Coordinator who truly has bought into the concept of patient involvement in the work of the ERN eases the path for patient engagement with the other clinicians who active in the network and in the working groups.
- This particular project had very enthusiastic clinical leads who also saw the need for developing these consensus statements. Working together in areas of common interest greatly facilitates patient engagement.
- 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).

## Annex 1

### The Postoperative management of patients with EA

The ePAG Advocates provided inputs on the different items listed below in the blue boxes.

#### Diagnostic procedures in patients with suspected esophageal atresia

	DIAGNOSTICS	Consensus	%	Votes	Median [range]
1	A nasogastric tube 10Fr or larger (modified for preterm infants) should be routinely inserted as a diagnostic procedure in cases with suspected EA.	+	100	15/15	9 [7-9]
2	A thoracoabdominal X-ray should be routinely performed as a preoperative diagnostic procedure.	+	100	15/15	9 [8-9]
3	An ultrasound of the abdomen (including kidney/urinary tract) should be routinely performed as a preoperative diagnostic procedure.	-	40	6/15	3 [1-9]
4	An ultrasound of the spine should be routinely performed as a preoperative diagnostic procedure.	-	6.7	1/15	1 [1-9]
5	Echocardiography should be routinely performed as a preoperative diagnostic procedure, especially regarding the exclusion of a right descending aorta.	+	100	15/15	9 [7-9]
6	A contrast-study of the upper esophageal pouch should be routinely performed as a preoperative diagnostic procedure.	-	13.3	2/15	1 [1-9]

Note: The patients didn't participate in the voting process however a lot of information for Patient representatives for counselling families were shared so it was important to answer those questions.

#### Preoperative management of patients with esophageal atresia\*

	PREOPERATIVE MANAGEMENT	Consensus	%	Votes	Median [range]
7	A repleg 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 the operation theatre

#### Operative management of patients with esophageal atresia

	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	+	100	15/15	9 [8-9]

	thoracotomy.				
22	Entry through the 4 <sup>th</sup> 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]
29	The thoracoscopic approach offers the advantage of faster recovery compared to the conventional approach.	-	53.3	8/15	6 [1-9]
30	The thoracoscopic approach offers the advantage of better cosmesis compared to the conventional approach.	+	94.1	16/17	9 [5-9]
31	The thoracoscopic approach offers the advantage of less musculoskeletal sequelae compared to the conventional approach.	+	86.7	13/15	8 [5-9]
32	The maximum insufflation pressure of CO <sub>2</sub> during thoracoscopy should not exceed 5 mmHg.	+	100	14/14	9 [6-9]
33	Maximum duration of thoracoscopic operation should be 3 hours.	+	92.9	13/14	8 [4-9]
34	The thoracoscopic approach has the disadvantage of longer operative time compared to the conventional approach.	+	92.9	13/14	7.5 [2-9]
35	The thoracoscopic approach has a negative pathophysiological impact (acidosis, cerebral oxygenation impairment) compared to the conventional approach.	-	30.8	4/13	5 [1-7]
36	The thoracoscopic approach has the disadvantage of a higher complication rate compared to the conventional approach.	-	35.7	5/14	3.5 [1-8]
37	A transanastomotic tube should be routinely inserted.	+	80	12/15	8 [1-9]
38	The tracheoesophageal fistula should preferably be closed by transfixing suture.	+	100	14/14	9 [6-9]
39	The anastomosis should be preferably performed with absorbable sutures.	+	85.7	12/14	8 [1-9]
40	The esophageal anastomosis should be preferably performed with interrupted sutures.	+	100	14/14	9 [6-9]
41	A chest drain should be routinely placed.	-	21.4	3/14	1 [1-9]
42	There is no place for routine fundoplication in patients with EA during the initial operation.	+	100	18/18	9 [8-9]

### Postoperative management of patients with esophageal atresia

	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]

## Annex 2

### Literature meeting the criteria of CEBM Level 1 Evidences

Statement	Domain	Reference	Study Type
The muscle-sparing approach is the recommended approach for conventional thoracotomy.	Operative Management	Askarpour S et al., Arq Bras Cir Dig 2018 [32]	RCT
The azygos vein should be preserved whenever possible.	Operative Management	Upadhyaya VD et al., Eur J Pediatr Surg. 2007 [33]	RCT
		Sharma S et al., Pediatr Surg Int 2007 [34]	RCT
The thoracoscopic approach has the disadvantage of a negative pathophysiological impact (acidosis, cerebral oxygenation impairment) compared to the conventional open approach.	Operative Management	Bishay M et al., Ann Surg 2013 [35]	Pilot RCT
An anastomotic leakage within the first 4 postoperative days may be considered for surgical revision.	Postoperative Management	Vaghela MM et al., J Pediatr Surg 2017 [36]	RCT

§ In accordance with the Oxford CEBM Levels of Evidence as published in 2009 [30]

RCT, randomized controlled trial

## Annex 3

### **Next step - 2nd ERNICA Consensus Conference on "Long-gap Esophageal Atresia: Perioperative, Surgical and Long-term Management"**

Email received on May 27<sup>th</sup> 2019 to all the participants, including the ePAG Advocates (so we patients are seen as colleagues!)

*Dear Colleague,*

*The 2nd ERNICA Consensus Conference on "Long-gap Esophageal Atresia: Perioperative, Surgical and Long-term Management" will be held on the 13th and 14th November 2019 in Berlin. We would like to get you involved in the preparation of this conference.*

*The organizing committee created a list including all potentially relevant aspects and points for discussion. The items were discussed in detail during the recent ERNICA meeting in Padua (11th - 12th April 2019) with all participants and were modified thereafter.*

*A RedCap (Research Electronic Data Capture) has been built up in order to manage an online survey among all participants. The aim of this survey is to prioritize each item on a 5 point Likert scale to obtain an order of the items according to their importance.*

*We kindly ask you to fill out the survey (please find the link below) by 15th June 2019.*

*Suggestions on any important topic that we might have missed are welcome.*

*After incorporating the feedback of all participants, the list will be made accessible in its final version including the prioritization.*

*Many thanks for your co-operation in advance.*

*Best regards,*