European Reference Network
for rare or low prevalence complex diseases

Network Inherited and Congenital Anomalies (ERNICA)

ERNICA CONSENSUS CONFERENCE ON THE PERIOPERATIVE, SURGICAL AND LONG-TERM MANAGEMENT OF PATIENTS WITH LONG-GAP ESOPHAGEAL ATRESIA

WITH EXPLANATIONS FOR PATIENTS AND FAMILIES

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INTRODUCTION

ERNICA is the **E**uropean **R**eference **N**etwork for Rare **I**nherited **C**ongenital **A**nomalies. This is a network of experts from hospitals across Europe that are specialised in the care of rare inherited congenital anomalies. Patient groups from across Europe are also involved. The aim of the network is to share disease-specific knowledge and expertise to ultimately improve the care of patients. More information about ERNICA can be found on the ERNICA website.

Esophageal Atresia (EA) is a rare birth defect where a part of the esophagus, the tube connecting the mouth to the stomach, is missing. Esophagael Atresia is an area of focus for ERNICA.

In 2019, a second ERNICA consensus conference took place. The aim of this conference was to establish consensus on the *Perioperative, Surgical and Long-term Management of Patients with Long-gap Esophageal Atresia*, by reviewing the latest scientific evidence and considering expert opinion.

24 ERNICA representatives attended from 9 European countries, including pediatric surgeons, pediatric gastroenterologists, a neonatologist, a pulmonologist, a methodologist and patient representatives. An anonymous online voting process took place and consensus was reached on 90 out of 99 items (91%). A scientific paper which reports the findings was published in The European Journal of Pediatric Surgery in 2020.

This document seeks to summarise the outcome of the consensus conference for patients and families. The consensus statements are listed in their relevant sections and it is noted whether consensus was reached, or not.

\+ = means 75% + of the experts/attendees involved agreed with the statement
\- = means that the experts/attendees disagreed with this statement (less than 75% agreed)

Additional explanations for patients and parents are also provided for items that are considered particularly important to understand.

TOFS (Tracheo-Oesophageal Fistula Support) is a UK-based charity dedicated to improving the lives of people born with Esophageal Atresia and tracheoesophageal fistula (and their carers). Tracheoesophageal fistula is when a child is born with an abnormal connection between the trachea (also known as the 'windpipe') and the esophagus. Most children born with tracheoesophageal fistula also have Esophageal Atresia. TOFS has developed a glossary of terms which you might find useful to look to alongside this document. This glossary is available in various languages. **Note:** As a UK-based charity, TOFS uses English spellings for its (English) materials E.g., *Esophageal Atresia is spelt* → *Oesophageal Atresia*

Note: *The contents of this document are based on a translation of the summary developed by KEKS, the German Esophageal Atresia Patient Support Group in 2019. You can access the German version via their website: https://www.keks.org/keks-hilft/projekte/ernica-leitlinien/*

How is esophageal atresia (EA) diagnosed and how are other problems clarified after birth?

	Statement	Consensus/no consensus	Explanations for patients/parents
1	A nasogastric tube 10 Fr or larger (modified for preterm infants) should be routinely inserted as a diagnostic procedure in cases with suspected esophageal atresia.	+	If esophageal atresia is suspected, a long, thin tube called a nasogastric tube is passed through the patient's nose or mouth into the esophagus for further investigation.
2	A thoracoabdominal X-ray should be routinely performed.	+	
3	An ultrasound of the abdomen (including kidney/urinary tract) should be routinely performed within the first week of life.	+	An ultrasound of the kidneys and other abdominal organs should be done within the first week of life. An ultrasound is a way of taking images inside the body.
4	Echocardiography should be routinely performed, especially to exclude a right descending aorta.	+	An examination of the heart should be done, in particular to rule out further malformations of the large blood vessels. An examination of the heart and in particular the position of the aorta (the main artery that carries blood away from the heart to the rest of the body) is very important for the planning of the operation.
5	A contrast study of a potential upper esophageal pouch should be routinely performed as a preoperative diagnostic procedure.	-	The upper blind end of the esophagus should not be exposed with a contrast liquid because of the risk of aspiration, which is fluid entering the lungs.
6	Gap measurement by bougies via the upper and lower bougies/gastroscope via the lower pouch is a viable option.	+	
7	Gap measurement by contrast study (upper and lower pouches) is a viable option.	-	The gap should not be measured with a contrast liquid.
8	A bougie in the upper pouch at the time of tracheoscopy for distil fistula is a viable option for gap measurement	+	

WHAT IS LONG-GAP ESOPHAGEAL ATRESIA (EA)?

	STATEMENT	CONSENSUS/NO CONSENSUS	EXPLANATIONS FOR PATIENTS/PARENTS
1	ANY ESOPHAGEAL ATRESIA WITHOUT AIR IN THE ABDOMEN SHOULD BE ASSUMED TO BE A LONG GAP.	+	
2	ONLY PATIENTS WITH ESOPHAGEAL ATRESIA GROSS TYPES A AND B SHOULD BE CONSIDERED AS LONG GAP.	-	
3	PATIENTS WITH A DISTAL TRACHEOESOPHAGEAL FISTULA AT THE CARINA OR BELOW SHOULD BE CONSIDERED AS LONG GAP.	-	
4	ANY ESOPHAGEAL ATRESIA WITH A GAP OF THREE VERTEBRAL BODIES OR MORE SHOULD BE CONSIDERED AS LONG-GAP.	+	

How is esophageal atresia (EA) treated surgically?

	Statement	Consensus/no consensus	Explanations for patients/parents
A. Initial management before reconstruction			
1	A replogle tube should be routinely placed into the upper esophageal pouch to allow continuous low-pressure suction.	+	A replogle tube is a special tube that is placed through the mouth or nose into the esophagus. Through this tube all fluids are sucked out. This prevents leakage into the airway and therefore prevents fluid entering the lungs. A replogle tube is designed to extract saliva at low-pressure from the upper esophageal pouch.
2	Gastrostomy should usually be performed initially to allow enteral feeding and to stimulate growing of the stomach.	+	The placement of a feeding tube through the abdominal wall to the stomach allows the child to receive nutrition and stimulates the growth of the stomach in the first instance.
3	Formation of a cervical esophagostomy should be avoided.	+	A diversion of the upper esophageal pouch at the neck (salivary fistula) should be avoided.
4	Bougienage of the proximal and/or distal stump to enable delayed primary anastomosis should be avoided.	+	Bougienage or probing of the esophageal stump in order to lengthen it for a primary anastomosis should be avoided.
5	Tracheobronchoscopy under spontaneous breathing should be performed in all patients.	+	Tracheobronchoscopy (MLB) under spontaneous breathing should be performed in all patients.
6	Tracheobronchoscopy should evaluate the presence of vocal cords, airway anomalies (e.g., cleft), proximal/distal fistula location, and tracheobronchomalacia.	+	Tracheoscopy (MLB) (assessment of the airway) to rule out an upper fistula or other malformations is mandatory in order to prevent avoidable difficulties during the operation.
7	Parents should be routinely informed during counseling about all different surgical options (replacement strategies, lengthening procedure, timing, minimal invasive, and conventional techniques).	+	Parents should be informed about all possible surgical procedures, as well as discussing the possibility of minimally invasive surgery. Minimally invasive surgery is surgery involving as little incision into the body as possible.
8	Parents should be able to request a second opinion and made aware of patient support organisations.	+	Parents should be given the opportunity for a second opinion, and should also be encouraged to contact the appropriate patient support group.
9	Pain assessment and management protocols should be applied.	+	Pain assessment and pain management protocols should be applied.
10	Parental involvement and training are an essential integral part of care.	+	Involving and training parents in caring for children is an important part of treatment.
11	Early oral stimulation, including sensory stimulation and sham feeding, is required to prevent abnormal oral feeding behaviour, especially in the case of delayed anastomosis.	+	Early oral stimulation and sham feeding are necessary to avoid feeding disorders, especially in the case of delayed anastomosis.

12	Sham feeding should be performed as soon as possible, including when a Replogle tube is in place.	+	Sham feeding should begin as early as possible, even if a Replogle tube is in place. Advice should be taken from the local team if any choking or breathing problems arise.
13	Professional nutritional assessment and support are mandatory to prevent undernutrition.	+	Professional advice on nutrition is essential to avoid under-nutrition.
B. Principle statements on esophageal reconstruction techniques			
14	Preserving the native esophagus should be preferred as initial management.	+	Keeping your child's own esophagus should initially be the preferred solution.
15	Delayed primary anastomosis should be preferred.	+	It is therefore preferable to wait and see (the two esophageal pouches very often still grow a little after the birth).
16	Axial lengthening procedure is a viable option.	+	
17	The Kimura advancement method of lengthening the upper pouch by extra-thoracic resiting a split fistula is not recommended.	+	The stretching of the esophageal tissue with the help of the ongoing surgical relocation of the saliva fistula (Kimura) is not recommended because of frequent complications.
18	Other esophageal lengthening techniques (flap, Livaditis circular myotomy, gastric division) are not recommended.	+	
19	Jejunal interposition is a viable option for esophageal replacement.	+	
20	Colonic interposition is a viable option for esophageal replacement.	-	
21	Gastric transposition is a viable option for esophageal replacement.	+	
22	Reconstructive surgery should only be performed in centrses with recognised expertise.	+	The reconstruction of the esophagus should definitely only be carried out in a designated expert centre.
C. Timing of esophageal reconstruction			
23	Gap assessment should be performed at 4–6 weeks.	+	
24	Delayed primary anastomosis should be performed at the age of around 2–3 months also depending on the gap assessment.	+	The delayed anastomosis should be carried out after 2-3 months at the earliest (depending on the distance between the ends [gap distance]).
25	Esophageal replacement should be performed at the age of 2–3 months also depending on the gap assessment	+	Esophageal replacement should be carried out after 2-3 months at the earliest (depending on the distance between the ends [gap distance]).
26	Reconstruction at a very early age is a viable option when esophageal lengthening technique is used.	+	

D. General aspects of operative management			
27	Antibiotics should be routinely administered perioperatively.	+	
28	A central venous line should be placed before the operation.	+	
29	An arterial line should be placed before the operation.	+	
E. Delayed primary anastomosis			
30	Horizontal or vertical or U-shaped (Bianchi) approaches (skin incision) are viable approaches for conventional thoracotomy.	+	
31	Muscle-sparing approach is the recommended approach for conventional thoracotomy.	+	
32	Entry through the fourth intercostal space is the recommended approach for conventional thoracotomy depending on assessment of gap length.	+	
33	The extrapleural approach is the preferred approach for thoracotomy.	+	
34	In cases with suspected right descending aorta, a right-sided thoracic approach is the first option.	+	
35	The azygos vein should be preserved whenever possible.	+	
36	The esophageal anastomosis should be preferably performed with absorbable sutures.	+	
37	The esophageal anastomosis should be preferably performed with interrupted sutures.	+	
38	A transanastomotic tube should be routinely inserted.	+	
39	A chest drain should be routinely placed.	-	
40	The thoracoscopic approach is a viable option.	+	Thorascopic (minimally invasive) surgery is a possible option.Thorascopic surgery is when a camera and instruments are used through small cuts in the chest to perform surgey (keyhole surgery).
41	The thoracoscopic approach should be only performed if suitable expertise is available.	+	Thorascopic (minimally invasive) surgery should only be sought if the necessary expertise is available. In addition to the pediatric surgeon, there should also be a pediatric anaesthetist with the appropriate experience.

F. LENGTHENING TECHNIQUES			
42	THORACOSCOPIC POUCH MOBILISATION AND PLACEMENT OF TRACTION SUTURES ARE A NOVEL TECHNIQUE THAT SHOWS PROMISE, BUT SHOULD ONLY BE PERFORMED IN SPECIALISED CENTRES WITH PROSPECTIVE REVIEW AND REPORTING OF OUTCOMES.	+	
43	OPEN POUCH MOBILISATION AND PLACEMENT OF TRACTION SUTURES ARE A VIABLE TECHNIQUE THAT SHOULD ONLY BE PERFORMED IN SPECIALISED CENTRES WITH PROSPECTIVE REVIEW AND REPORTING OF OUTCOMES.	+	

How, if necessary, is the esophagus replaced?

	Statement	Consensus/no consensus	Explanations for patients/parents
Gastric transposition			
1	The anastomosis for gastric transposition should be routinely performed on the patient's right side.	+	
2	Thoracotomy for gastric transposition should be avoided whenever possible.	+	
3	Partial gastric transposition with intrathoracic anastomosis should be avoided.	+	A partial stomach 'pull-up' generally results in multiple complications, and is therefore not a proven treatment strategy
4	A pyloroplasty (Mikulicz) should be routinely performed.	+	
5	Laparoscopically assisted gastric transposition is a viable option.	+	
6	A jejunostomy should be routinely performed to allow postoperative feeding unless sham feeding is well established.	+	A jejunal tube should be routinely placed so that learning to eat can be separated from growth/development (less stress).
Jejunal Interposition			
7	A cervical esophagostomy is a contraindication for jejunal interposition.	+	
Colonic Interposition			
8	The right hemicolon should be routinely used in an isoperistaltic manner.	+	
9	The preferred position is the posterior mediastinum.	+	

How will the child be cared for after the operation?

	Statement	Consensus/no consensus	Explanations for patients/parents
1	Postoperative ventilation and relaxation should be performed for up to 5 days in anastomoses under tension.	+	
2	Routine postoperative antibiotic prophylaxis beyond 48 hrs is not recommended.	+	
3	A postoperative contrast study of the esophagus should be routinely performed before the initiation of oral feeding.	-	
4	Enteral feeding should be routinely initiated on the second postoperative day via a gastric or jejunal route.	+	
5	A clinical checklist should be made available including items which should be performed before fiRST discharge (e.g., abdominal and renal ultrasound, resuscitation training for parents/caregivers).	+	
6	Resuscitation training for parents and caregivers is mandatory before discharge.	+	Emergency training for parents and other carers is absolutely necessary before discharge from the hospital

What should (at a minimum) good aftercare look like?

	Statement	Consensus/no consensus	Explanations for patients/parents
1	There should be a structured schedule for lifelong follow-up.	+	Lifelong structured follow-up care is necessary.
2	There should be an interdisciplinary follow-up program including surgeons, gastroenterologists, pulmonologists, otolaryngologists, nutrition counseling and others, with one specialist leading.	+	The follow-up care should be multi-disciplinary, but requires an EA expert who provides overall coordination
3	Proton pump inhibitors (PPIs) should be used for antacid prophylaxis.	+	PPI (medication that is swallowed to treat reflux) should be given as a preventive measure

4	Antacid medication should be routinely administered to at least until the age of 12 months.	+	The PPIs should be given routinely for the entire first year of life.
5	Antacid therapy should be tapered at the end of prophylaxis.	+	When the initial preventive treatment for reflux is stopping this should be slowly reduced.
6	In patients with symptoms, anastomotic strictures should be diagnosed by contrast and/or endoscopy.	+	
7	Anastomotic stricture should be managed by balloon or semirigid dilatation.	+	
8	The definition of recurrent anastomotic stricture is 3 anastomotic stricture relapses requiring dilatation.	+	A stenosis/stricture or narrowing at the anastomosis that requires at least three dilatations is considered a complicated stenosis. Complicated stenoses should only be treated in centres with proven expertise.
9	Topical application of mitomycin C is a viable option in patients with recurrent strictures.	+	
10	Intralesional steroids are a viable option in patients with recurrent strictures.	+	
11	Stents are a viable option in patients with recurrent strictures, but should only be used with caution.	+	
12	Outpatient clinical and nutritional assessment should be performed every 3 months during the first year after reconstruction.	+	After discharge from the hospital, the child's nutritional condition (and feeding/eating drinking) should be discussed every 3 months in the first year of life as part of a follow-up appointment
13	Upper GI endoscopy and/or pH–impedance-metry should be performed 1 year after reconstruction after tapering proton pump inhibitors.	+	After the PPI medication has been reduced, control EGD (endoscopy) and / or pH-impedence study (24hour reflux testing), should be carried out at around 1 year of age, even if no symptoms have occurred. A barium swallow does not replace these controls.
14	Outpatient clinical and nutritional assessment is recommended at least every second year until transition.	+	Until transition (transition from child toadult healthcare), nutrition and quality of life * should also be discussed every 2 years as part of the follow-up appointment (*age-appropriate, balanced, adequate/appropriate).
15	At least two additional endoscopies of the upper gastrointestinal tract should be performed until transition.	+	At least two other endoscopies (unless some special reason exists) should be carried out as a check-up until adulthood.
16	Respiratory review by pulmonologists should be routinely performed for children and adolescents according to a specific schedule.	+	Lung function should be checked routinely and regularly according to a specific schedule. (e.g. every two years).
17	Contrast study of the upper gastrointestinal tract should not be routinely used for monitoring children and adolescents according to a specific schedule.	+	A barium swallow is not recommended as a routine check unless there are particular symptoms.

18	Bronchoscopy is recommended for symptomatic children.	+	
19	Adult patients need surveillance as per ESPGHAN guidelines: (1) routine endoscopy every 5–10 years, (2) endoscopy if new or worsening symptoms occur, and (3) in presence of Barrett as per consensus recommendations.	+	Adults should have an endoscopy routinely every 5 to 10 years even without symptoms. The ESPGHAN guidelines must be observed in the event of new or worsening symptoms and/or a diagnosis of Barrett's esophagus.
20	A specific transition program for adolescents with long-gap esophageal atresia should be organised.	+	A special transition program (to support chidren move to adult care) should be organised by the clinic for adolescents with repaired long-gap EA.
21	Quality of life assessment using a validated instrument should be offered during follow-up in children, adolescents, and adult patients.	+	An assessment of quality of life should be offered for children, adolescents and adults as part of aftercare.
22	Screening for dumping syndrome in children is required especially in children with microgastria, or when pyloroplasty or antireflux surgery has been performed.	+	An examination of children for dumping syndrome is particularly necessary in the presence of a microgastria (very small stomach), after a pyloroplasty (surgical widening of the outlet of the stomach) or anti-reflux surgery (fundoplication).
23	When endoscopy is performed, there should be awareness of eosinophilic esophagitis, and biopsies should be taken according to ESPGHAN guidelines.	+	During an endoscopy of the esophagus, tissue samples known as biopsies should always be taken, e.g. to rule out eosinophilic esophagitis (EoE).

WHAT SHOULD ALWAYS BE VALID IN PRINCIPLE?

	STATEMENT	CONSENSUS/NO CONSENSUS	EXPLANATIONS FOR PATIENTS/PARENTS
1	WHEN THE DIAGNOSIS OF LONG-GAP ESOPHAGEAL ATRESIA IS CONFIRMED, THE PATIENT SHOULD BE REFERRED TO A CENTRE OF EXPERTISE IN ESOPHAGEAL RECONSTRUCTIVE SURGERY.	+	A PREGNANT WOMAN SUSPECTED OF HAVING A BABY WITH A DIAGNOSIS OF ESOPHAGEAL ATRESIA SHOULD BE REFERRED TO A SPECIALISED CENTRE WITH THE APPROPRIATE NECESSARY STRUCTURE AND NUMBER OF CASES.
2	LONG-GAP ESOPHAGEAL ATRESIA SHOULD BE MANAGED IN CENTERS WITH EXPERTISE IN ESOPHAGEAL RECONSTRUCTIVE SURGERY, PREFERABLY WITH MORE THAN TWO CASES PER YEAR.	+	A CHILD WITH LONG-GAP ESOPHAGEAL ATRESIA SHOULD ONLY BE TREATED IN A SPECIALISED CENTRE THAT HAS THE APPROPRIATE NECESSARY STRUCTURE AND NUMBER OF CASES.
3	WHEN LONG-GAP ESOPHAGEAL ATRESIA IS SUSPECTED, REFERRAL TO ANTENATAL MULTIDISCIPLINARY COUNSELING IN A CENTRE OF EXPERTISE SHOULD BE MADE.	+	IF A LONG-GAP ESOPHAGEAL ATRESIA IS SUSPECTED, THE PARENTS-TO-BE SHOULD BE REFERRED TO A SPECIALISED CENTRE WITH PROVEN EXPERTISE FOR MULTIDISCIPLINARY ADVICE. SUCH CENTRES SHOULD HAVE THE APPROPRIATE NECESSARY STRUCTURE AND NUMBER OF CASES.
4	PARENTS OF ESOPHAGEAL ATRESIA PATIENTS SHOULD BE INFORMED ABOUT, AND ENCOURAGED TO CONTACT PARENT AND PATIENT SUPPORT GROUPS AS EARLY AS POSSIBLE.	+	PARENTS SHOULD BE ENCOURAGED TO CONTACT THEIR NATIONAL (OR MOST APPROPRIATE BY LANGUAGE OR GEOGRAPHY) PARENT/PATIENT SUPPORT GROUPS AS EARLY AS POSSIBLE.

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