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ERNICA Consensus Conference on the Management of Patients with Long-Gap Esophageal Atresia: Perioperative, Surgical, and Long-Term Management

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Abstract

Introduction Evidence supporting best practice for long-gap esophageal atresia is limited. The European Reference Network for Rare Inherited Congenital Anomalies (ERNICA) organized a consensus conference on the management of patients with long-gap esophageal atresia based on expert opinion referring to the latest literature aiming to provide clear and uniform statements in this respect.

Materials and Methods Twenty-four ERNICA representatives from nine European countries participated. The conference was prepared by item generation, item prioritization by online survey, formulation of a final list containing items on perioperative, surgical, and long-term management, and literature review. The 2-day conference was held in Berlin in November 2019. Anonymous voting was conducted via an

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internet-based system using a 1 to 9 scale. Consensus was defined as ≥75% of those voting scoring 6 to 9.

Results Ninety-seven items were generated. Complete consensus (100%) was achieved on 56 items (58%), e.g., avoidance of a cervical esophagostomy, promotion of sham feeding, details of delayed anastomosis, thoracoscopic pouch mobilization and placement of traction sutures as novel technique, replacement techniques, and follow-up. Consensus ≥75% was achieved on 90 items (93%), e.g., definition of long gap, routine pyloroplasty in gastric transposition, and avoidance of preoperative bougienage to enable delayed anastomosis. Nineteen items (20%), e.g., methods of gap measurement were discussed controversially (range 1–9).

Conclusion This is the first consensus conference on the perioperative, surgical, and long-term management of patients with long-gap esophageal atresia. Substantial statements regarding esophageal reconstruction or replacement and follow-up were formulated which may contribute to improve patient care.

Introduction

In 1 out of 10 patients with esophageal atresia, primary anastomosis of the two esophageal ends is not feasible[1,2] which poses an additional major challenge to restore continuity.[1–7] The rarity of the condition, the variability in case definition, multiple approaches to management and follow-up, and the heterogeneity of the reported outcomes contribute to this challenge.[3] In addition, evidence supporting best practice for long-gap esophageal atresia is limited, mostly consisting of single-center retrospective reviews with low numbers of patients, and low quality of data to date.[1–5] As a result, there is a variety of coexisting protocols on the perioperative, surgical, and long-term management of patients with *long-gap* esophageal atresia based on opinion rather than on evidence. This was confirmed in a recent survey of pediatric surgeons demonstrating the variability in opinion, in terms of both case definition and preferred operative approaches.[8]

The European Reference Network for Rare Inherited Congenital Anomalies (ERNICA) has been established in response to the European Commission's call for the setup of European Reference Networks for rare diseases in 2017.[9] The network seeks to deliver high standards of care, to build capacity where there is lack of knowledge and infrastructure, and to promote optimal patient care for rare inherited and congenital digestive tract-related disorders from pediatric age to adulthood.[10–13]

ERNICA organized a first consensus conference on the pre-, peri-, and postoperative management as well as on the follow-up of patients with esophageal atresia with tracheoesophageal fistula in October 2018.[14,15] ERNICA has now conducted a second consensus conference which focused on the management of patients with long-gap esophageal atresia based on expert opinions referring to the latest literature. The aim of this conference was to develop clear and uniform statements in this respect.

Materials and Methods

The general methodological approach has been recently published when presenting the results of the first ERNICA consensus conference.[14,15] It consisted of two parts: (1) diagnostics, preoperative, operative, and postoperative management[14] and (2) follow-up and framework.[15]

The conference dealt exclusively with the management of patients with *long-gap* esophageal atresia, and took place in Berlin on November 13–14, 2019. In total, 24 ERNICA representatives from nine European countries participated: 16 pediatric surgeons, 2 pediatric gastroenterologists, 1 neonatologist, 1 pediatric pulmonologist, 3 representatives of patient support groups acting under the umbrella of the Federation of Esophageal Atresia and Tracheo-Esophageal Fistula Support Groups (EAT).[16] One nonsurgeon methodologist (S.E.) took part in all steps of the preparation and the conference itself.

With regard to literature selection, publications with the highest grade of evidence according to the Centre for Evidence-Based Medicine (CEBM) classification were suggested to be preferred[17] as previously reported.[14,15] Literature was distributed and made available to all participants via a Dropbox (Dropbox Inc., San Francisco, California, United States, 2007) link prior to the conference.

The preparation and implementation of the conference included the following steps[14,15]: (1) generation of a list of items; (2) prioritization of the items using the online REDCap electronic data capture tools[18]; (3) literature-based discussion of all items on the perioperative, surgical, and long-term management of patients with long-gap esophageal atresia during the conference, formulation of statements; (4) anonymous voting via the internet-based system VoxVote (VoxVote, Breda, The Netherlands) using a 1 to 9 scale.[19]

It was suggested to the participants that they abstained from voting on any individual item when they felt that they had no expertise or an opinion on that item. Therefore, participants were allowed to vote online "no relevant expertise on this statement." As a result, the number of scoring participants varied for individual statements. Consensus was defined as ≥75% of those voting scored 6, 7, 8, or 9, excluding those who declared no relevant expertise on that statement.[14,15]

The wordings of the statements on items were updated during the discussion by the participants of the conference

and prepared for voting by the nonsurgical methodologist (S. E.) who did not himself vote.

The final scores and the consensus results were shown to all participants after all votes were obtained, but individual scores remained anonymous. Details of the discussions, in particular the controversial aspects, were documented throughout the conference by C.D.

Results and Consensus Statements

Item Generation and Prioritization

The systematic literature search and the discussion of the members of the ERNICA Workstream *Congenital Malformations and Diseases of the Esophagus* during the third ERNICA Annual Meeting in Padua, Italy on April 11–12, 2019, resulted in a total of 87 items.

After the online prioritization phase, two items were excluded. Following the participants' suggestions, one new item was added. Consequently, the list included 86 items prior to the conference, for which literature was obtained and circulated.

As a result of the presentations by the domain leaders and active discussion during the conference, 14 items were excluded, and 25 were added as some items were split into several separate questions.

Finally, 97 items were reworded and confirmed for voting. This included 8 items in the domain *Diagnostics*, 4 items in the domain *Definitions*, 43 items in the domain *Esophageal Reconstruction*, 9 items in the domain *Esophageal Replacement*, 6 items in the domain *Postoperative Management*, 23 items in the domain *Follow-up*, and 4 items in the domain *Framework* (►Tables 1–7).

Several items of the domain *Follow-up* have been adopted from the guidelines released by the *European Society for Pediatric Gastroenterology Hepatology and Nutrition* (ESPGHAN) and the *North American Society for Pediatric Gastroenterology, Hepatology and Nutrition*[20] (NASPGHAN) and were discussed from a surgical perspective. Some items had already been discussed in a different context during the first ERNICA consensus conference on the management of patients with esophageal atresia and tracheoesophageal fistula. Therefore, the wording might be the same or modified to some extent, but was considered essential to be rediscussed to determine whether the statement was also relevant to the group of patients with long-gap esophageal atresia.

Consensus

Detailed results on the consensus among participants of the conference are summarized in ►Tables 1 to 7. For 19 items (20%), the results ranged from 1 to 9 reflecting controversial opinions in these cases. In six of these (32%), no consensus was reached.

►Table 8 depicts the general distribution of voting results.

There was a deviation from the established voting process using the 1 to 9 scale for one statement voting on the preferred option for esophageal replacement. In this case, participants were asked to vote on only one single option. Nine (39%) participants preferred gastric transposition, 4 (17%) participants preferred jejunal interposition, and 3 (13%) participants preferred colonic interposition for esophageal replacement (►Fig. 1). For this vote, seven (30%) participants abstained from voting.

Controversial Items Discussed without Voting

Several items were discussed controversially, and the participants agreed verbally that more data from future research should be available before a meaningful question and vote

Table 1 Diagnostics

	Statement	Consensus	%	Votes	Median (range)
1[a]	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	+	100	21/21	9 (8–9)
2[a]	A thoracoabdominal X-ray should be routinely performed	+	100	21/21	9 (9–9)
3[b]	An ultrasound of the abdomen (including kidney/urinary tract) should be routinely performed within the first week of life	+	100	20/20	9 (7–9)
4[a]	Echocardiography should be routinely performed, especially to exclude a right descending aorta	+	100	21/21	9 (9–9)
5[a]	A contrast study of a potential upper esophageal pouch should be routinely performed as a preoperative diagnostic procedure	−	16.7	3/18	2 (1–9)
6	Gap measurement by bougies via the upper and lower bougies/gastroscope via the lower pouch is a viable option	+	94.7	18/19	9 [5–9]
7	Gap measurement by contrast study (upper and lower pouches) is a viable option	−	40	8/20	3 (1–9)
8	A bougie in the upper pouch at the time of tracheoscopy for distal fistula is viable option for gap measurement	+	83.3	15/18	8 (2–9)

Abbreviation: ERNICA, European Reference Network for Rare Inherited Congenital Anomalies.

[a]This item has already been discussed during the first ERNICA consensus conference on the management of patient with esophageal atresia with tracheoesophageal fistula.

[b]This item has already been discussed during the first ERNICA consensus conference on the management of patient with esophageal atresia with tracheoesophageal fistula, but in another context.

Table 2 Definitions

	Statement	Consensus	%	Votes	Median (range)
1	Any esophageal atresia without air in the abdomen should be assumed to be a long gap	+	100	22/22	9 (7–9)
2	Only patients with esophageal atresia Gross types A and B should be considered as long gap	–	42.9	9/21	5 (1–9)
3	Patients with a distal tracheoesophageal fistula at the carina or below should be considered as long gap	–	54.5	12/22	6.5 (1–9)
4	Any esophageal atresia with a gap of three vertebral bodies or more should be considered as long-gap	+	76.2	16/21	8 (1–9)

Table 3 Esophageal reconstruction

	Statement	Consensus	%	Votes	Median (range)
a. Initial management before reconstruction					
1[a]	A replogle tube should be routinely placed into the upper esophageal pouch to allow continuous low pressure suction	+	100	22/22	9 (6–9)
2	Gastrostomy should usually be performed initially to allow enteral feeding and to stimulate growing of the stomach	+	100	22/22	9 (6–9)
3	Formation of a cervical esophagostomy should be avoided	+	100	21/21	9 (8–9)
4	Bougienage of the proximal and/or distal stump to enable delayed primary anastomosis should be avoided	+	78.9	15/19	9 (2–9)
5	Tracheobronchoscopy under spontaneous breathing should be performed in all patients	+	100	22/22	9 (8–9)
6	Tracheobronchoscopy should evaluate the presence of vocal cords, airway anomalies (e.g., cleft), proximal/distal fistula location, and tracheobronchomalacia	+	100	23/23	9 (9–9)
7	Parents should be routinely informed during counseling about all different surgical options (replacement strategies, lengthening procedure, timing, minimal invasive, and conventional techniques)	+	81.8	18/22	9 (1–9)
8	Parents should be able to request a second opinion and made aware of patient support organizations	+	100	23/23	9 (6–9)
9	Pain assessment and management protocols should be applied	+	100	23/23	9 (7–9)
10	Parental involvement and training are an essential integral part of care	+	100	23/23	9 (8–9)
11	Early oral stimulation, including sensory stimulation and sham feeding, is required to prevent abnormal oral feeding behavior, especially in the case of delayed anastomosis	+	100	23/23	9 (7–9)
12	Sham feeding should be performed as soon as possible, including when a Replogle tube is in place	+	100	23/23	9 (6–9)
13	Professional nutritional assessment and support are mandatory to prevent undernutrition	+	100	23/23	9 (6–9)
b. Principle statements on esophageal reconstruction techniques					
14	Preserving the native esophagus should be preferred as initial management	+	100	22/22	9 (9–9)
15	Delayed primary anastomosis should be preferred	+	100	21/21	9 (6–9)
16	Axial lengthening procedure is a viable option	+	83.3	15/18	7 (3–9)
17	The Kimura advancement method of lengthening the upper pouch by extra-thoracic resiting a spit fistula is not recommended	+	100	17/17	9 (6–9)
18	Other esophageal lengthening techniques (flap, Livaditis circular myotomy, gastric division) are not recommended	+	88.2	15/17	9 (2–9)
19	Jejunal interposition is a viable option for esophageal replacement	+	87.5	14/16	8.5 (2–9)
20	Colonic interposition is a viable option for esophageal replacement	–	47.1	8/17	5 (1–9)
21	Gastric transposition is a viable option for esophageal replacement	+	100	17/17	9 (6–9)
22		+	100	22/22	9 (6–9)

(*Continued*)

Table 3 (*Continued*)

	Statement	Consensus	%	Votes	Median (range)
	Reconstructive surgery should only be performed in centers with recognized expertise				
c. Timing of esophageal reconstruction					
23	Gap assessment should be performed at 4–6 weeks	+	100	19/19	9 (6–9)
24	Delayed primary anastomosis should be performed at the age of around 2–3 months also depending on the gap assessment	+	100	18/18	9 (7–9)
25	Esophageal replacement should be performed at the age of 2–3 months also depending on the gap assessment	+	82.4	14/17	8 (1–9)
26	Reconstruction at a very early age is a viable option when esophageal lengthening technique is used	+	93.8	15/16	8 (5–9)
d. General aspects of operative management					
27[a]	Antibiotics should be routinely administered perioperatively	+	100	20/20	9 (6–9)
28[a]	A central venous line should be placed before the operation	+	100	20/20	9 (6–9)
29[a]	An arterial line should be placed before the operation	+	94.4	17/18	9 (1–9)
e. Delayed primary anastomosis					
30[a]	Horizontal or vertical or U-shaped (Bianchi) approaches (skin incision) are viable approaches for conventional thoracotomy	+	100	15/15	9 (7–9)
31[a]	Muscle-sparing approach is the recommended approach for conventional thoracotomy	+	100	17/17	9 (7–9)
32[a]	Entry through the fourth intercostal space is the recommended approach for conventional thoracotomy depending on assessment of gap length	+	100	16/16	9 (7–9)
33[a]	The extrapleural approach is the preferred approach for thoracotomy	+	86.7	13/15	9 (1–9)
34[a]	In cases with suspected right descending aorta, a right-sided thoracic approach is the first option	+	93.8	15/16	8 (1–9)
35[b]	The azygos vein should be preserved whenever possible	+	88.2	15/17	7 (4–9)
36[a]	The esophageal anastomosis should be preferably performed with absorbable sutures	+	88.2	15/17	9 (1–9)
37[a]	The esophageal anastomosis should be preferably performed with interrupted sutures	+	100	16/16	9 (7–9)
38[a]	A transanastomotic tube should be routinely inserted.	+	94.7	18/19	9 (5–9)
39[a]	A chest drain should be routinely placed	−	64.7	11/17	7 (2–9)
40[a]	The thoracoscopic approach is a viable option	+	100	16/16	9 (6–9)
41[a]	The thoracoscopic approach should be only performed if suitable expertise is available	+	95.2	20/21	9 (1–9)
f. Lengthening techniques					
42	Thoracoscopic pouch mobilization and placement of traction sutures are a novel technique that shows promise, but should only be performed in specialized centers with prospective review and reporting of outcomes	+	100	23/23	9 (6–9)
43	Open pouch mobilization and placement of traction sutures are a viable technique that should only be performed in specialized centers with prospective review and reporting of outcomes	+	86.4	19/22	9 (2–9)

Abbreviation: ERNICA, European Reference Network for Rare Inherited Congenital Anomalies.
[a]This item has already been discussed during the first ERNICA consensus conference on the management of patient with esophageal atresia with tracheoesophageal fistula.
[b]This item has already been discussed during the first ERNICA consensus conference on the management of patient with esophageal atresia with tracheoesophageal fistula, but consensus could not be reached on this item.

could be conducted. The discussion included in particular the following aspects:

Definitions

The participants agreed to define long-gap esophageal atresia as "any esophageal atresia without air in the abdomen" or "any esophageal atresia with a gap of three vertebral bodies or more" as the lowest common denominator. However, there was a debate on whether only patients with esophageal atresia Gross types A and B or also patients with a distal tracheoesophageal fistula at the carina or below should be considered as long-gap esophageal atresia. The participants voted on the two

Table 4 Esophageal replacement

	Statement	Consensus	%	Votes	Median (range)
Gastric transposition					
1	The anastomosis for gastric transposition should be routinely performed on the patient's right side	+	90.9	10/11	8 (5–9)
2	Thoracotomy for gastric transposition should be avoided whenever possible	+	91.7	11/12	8.5 (4–9)
3	Partial gastric transposition with intrathoracic anastomosis should be avoided	+	100	13/13	8 (6–9)
4	A pyloroplasty (Mikulicz) should be routinely performed	+	91.7	11/12	8 (1–9)
5	Laparoscopically assisted gastric transposition is a viable option	+	100	17/17	9 (6–9)
6	A jejunostomy should be routinely performed to allow postoperative feeding unless sham feeding is well established	+	95	19/20	8 (1–9)
Jejunal interposition					
7	A cervical esophagostomy is a contraindication for jejunal interposition	+	100	12/12	7 (6–)
Colonic interposition					
8	The right hemicolon should be routinely used in an isoperistaltic manner	+	100	10/10	8 (6–9)
9	The preferred position is the posterior mediastinum	+	100	12/12	8.5 (6–9)

Table 5 Postoperative management

	Statement	Consensus	%	Votes	Median (range)
1	Postoperative ventilation and relaxation should be performed for up to 5 d in anastomoses under tension	+	100	17/17	8 (6–9)
2	Routine postoperative antibiotic prophylaxis beyond 48 h is not recommended	+	95	19/20	9 (5–9)
3[a]	A postoperative contrast study of the esophagus should be routinely performed before the initiation of oral feeding	–	40	8/20	5 (1–9)
4	Enteral feeding should be routinely initiated on the second postoperative day via a gastric or jejunal route	+	85	17/20	8.5 (3–9)
5[a]	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)	+	95.5	21/22	9 (5–9)
6	Resuscitation training for parents and caregivers is mandatory before discharge	+	91.3	21/23	9 (1–9)

Abbreviation: ERNICA, European Reference Network for Rare Inherited Congenital Anomalies.
[a]This item has already been discussed during the first ERNICA consensus conference on the management of patient with esophageal atresia with tracheoesophageal fistula.

Table 6 Follow-up

	Statement	Consensus	%	Votes	Median (range)
1[a]	There should be a structured schedule for lifelong follow-up	+	100	23/23	9 (8–9)
2[a]	There should be an interdisciplinary follow-up program including surgeons, gastroenterologists, pulmonologists, otolaryngologists, nutrition counseling and others, with one specialist leading	+	100	23/23	9 (8–9)
3[a]	Proton pump inhibitors should be used for antacid prophylaxis	+	90	18/20	9 (3–9)
4[b]	Antacid medication should be routinely administered to at least until the age of 12 months	+	90.5	19/21	9 (2–9)
5[a]	Antacid therapy should be tapered at the end of prophylaxis	+	100	22/22	9 (6–9)
6[a]	In patients with symptoms, anastomotic strictures should be diagnosed by contrast and/or endoscopy	+	100	21/21	9 (8–9)
7[a]	Anastomotic stricture should be managed by balloon or semirigid dilatation	+	100	19/19	9 (8–9)
8[a]	The definition of recurrent anastomotic stricture is three anastomotic stricture relapses requiring dilatation	+	100	23/23	9 (7–9)

(*Continued*)

Table 6 (*Continued*)

	Statement	Consensus	%	Votes	Median (range)
9[b]	Topical application of mitomycin C is a viable option in patients with recurrent strictures	+	77.8	14/18	7.5 (1–9)
10[a]	Intralesional steroids are a viable option in patients with recurrent strictures	+	94.4	17/18	7.5 (5–9)
11	Stents are a viable option in patients with recurrent strictures, but should only be used with caution	+	94.7	18/19	9 (4–9)
12	Outpatient clinical and nutritional assessment should be performed every 3 month during the first year after reconstruction	+	100	23/23	9 (6–9)
13	Upper GI endoscopy and/or pH–impedance-metry should be performed 1 year after reconstruction after tapering proton pump inhibitors	+	100	19/19	9 (7–9)
14	Outpatient clinical and nutritional assessment is recommended at least every second year until transition	+	100	23/23	9 (7–9)
15[a]	At least two additional endoscopies of the upper gastrointestinal tract should be performed until transition	+	100	23/23	9 (6–9)
16	Respiratory review by pulmonologists should be routinely performed for children and adolescents according to a specific schedule	+	100	23/23	9 (7–9)
17	Contrast study of the upper gastrointestinal tract should not be routinely used for monitoring children and adolescents according to a specific schedule	+	100	23/23	9 (7–9)
18	Bronchoscopy is recommended for symptomatic children	+	100	23/23	9 (6–9)
19[a]	Adult patients need surveillance as per ESPGHAN guidelines: (1) routine endoscopy every 5–10 y, (2) endoscopy if new or worsening symptoms occur, and (3) in presence of Barrett as per consensus recommendations	+	100	23/23	9 (6–9)
20	A specific transition program for adolescents with long-gap esophageal atresia should be organized	+	100	23/23	9 (6–9)
21[a]	Quality of life assessment using a validated instrument should be offered during follow-up in children, adolescents, and adult patients	+	100	23/23	9 (6–9)
22	Screening for dumping syndrome in children is required especially in children with microgastria, or when pyloroplasty or antireflux surgery has been performed	+	95.7	22/23	9 (5–9)
23	When endoscopy is performed, there should be awareness of eosinophilic esophagitis, and biopsies should be taken according to ESPGHAN guidelines	+	100	22/22	9 (6–9)

Abbreviations: ERNICA, European Reference Network for Rare Inherited Congenital Anomalies; ESPGHAN, European Society for Pediatric Gastroenterology Hepatology and Nutrition; GI, gastrointestinal.
[a]This item has already been discussed during the first ERNICA consensus conference on the management of patient with esophageal atresia with tracheoesophageal fistula.
[b]This item has already been discussed during the first ERNICA consensus conference on the management of patient with esophageal atresia with tracheoesophageal fistula, but consensus could not be reached on this item.

Table 7 Framework

	Statement	Consensus	%	Votes	Median (range)
1	When the diagnosis of long-gap esophageal atresia is confirmed, the patient should be referred to a center of expertise in esophageal reconstructive surgery	+	100	23/23	9 (7–9)
2	Long-gap esophageal atresia should be managed in centers with expertise in esophageal reconstructive surgery, preferably with more than two cases per year	+	91.3	21/23	9 (1–9)
3	When long-gap esophageal atresia is suspected, referral to antenatal multidisciplinary counseling in a center of expertise should be made	+	95.7	22/23	9 (4–9)
4[a]	Parents of esophageal atresia patients should be informed about, and encouraged to contact parent and patient support groups as early as possible	+	100	23/23	9 (6–9)

Abbreviation: ERNICA, European Reference Network for Rare Inherited Congenital Anomalies.
[a]This item has already been discussed during the first ERNICA consensus conference on the management of patient with esophageal atresia with tracheoesophageal fistula.

Table 8 Consensus amongst participants of the second ERNICA consensus conference

Consensus	No. of items (%)
100%	56 (58)
>75%[a]	90 (93)
<75%	7 (7)

Abbreviation: ERNICA, European Reference Network for Rare Inherited Congenital Anomalies.
[a]Votes that scored 6, 7, 8, or 9 also including items that reached 100%.

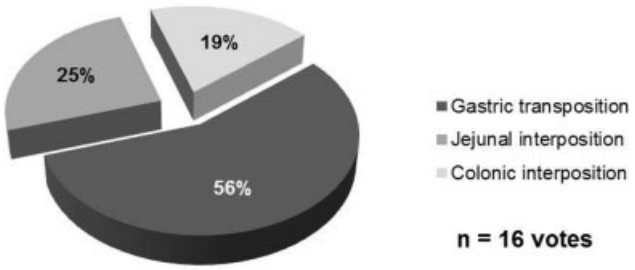

Fig. 1 Preferred option of esophageal replacement.

latter statements, though they did not reach consensus (►Table 2). In addition, there was agreement that "long gap" is not an appropriate term to define the condition of long-gap esophageal atresia in its entirety. The term "wide-gap" esophageal atresia, which is used in some countries to identify cases where the anastomosis cannot be achieved despite the presence of a fistula, was proposed to be introduced as an adequate definition. However, participants decided not to vote on this new definition to avoid more confusion as long-gap esophageal atresia is a well-established and accepted term.

Esophageal Reconstruction

It had been suggested to vote on the surgical option of a gastric tube for esophageal reconstruction. After extensive discussion, it was decided not to vote on this item due to limited evidence in the literature[21,22] and lack of personal experience within the group.

Postoperative Management

An attempt was made to formulate a statement on the commencement of oral feeding. However, most participants felt that a definition of a specific time point would not be appropriate as individual parameters, such as duration of ventilator dependency, have a decisive impact on the postoperative time management. Therefore, no statement was formulated on the commencement of oral feeding.

Follow-up

Complete consensus was reached on the management of anastomotic strictures with balloon or semirigid dilatation (►Table 6). In this context, participants discussed the indication of peri-interventional antibiotic prophylaxis. Even if most of the participants supported peri-interventional antibiotic prophylaxis, the evidence in the available literature was considered to be too low allowing a meaningful vote.

Participants voted on treatment options for recurrent anastomotic strictures, such as topical application of mitomycin C, intralesional steroids, and stents (►Table 6). Participants also considered whether to vote on the application of indwelling balloon dilatation or endoscopic knife for recurrent anastomotic strictures. Moreover, surgical resection and esophageal reanastomosis was proposed as viable option in cases of failed treatment. Nonetheless, evidence for each of these options is scarce,[23–25] and therefore, it was decided to abstain from formulating items for voting.

Complete consensus was reached on the statement that antacid therapy should be tapered at the end of prophylaxis (►Table 6). However, it was not possible to formulate a concrete instruction of how to taper the antacid medication due to a lack of evidence in literature and multiple suggestions by the participants.

Need for Further Research

The items on which either no consensus was reached or it was felt that there was insufficient evidence were suggested to be priorities for future research. ►Table 9 summarizes relevant topics which urgently need further studies.

Discussion

The management of long-gap esophageal atresia remains challenging with limited evidence and consensus on the definition, evaluation, and surgical approach.[2–4,9,26–28] The variety of management strategies that have been employed testify to the challenge it presents.[8]

In 2017, the International Network of Esophageal Atresia (INoEA) presented a position paper on the definition of long-gap esophageal atresia and the best diagnostic and treatment strategies also highlighting the necessity of experience and communication in the management of these challenging patients.[5] Recently, the American Pediatric Surgery Association (APSA) released a systematic review and 18 evidence-based guidelines, primarily based on levels 4 and 5 evidence, on the management of long-gap esophageal atresia.[3]

We hereby present the results of the second ERNICA consensus conference focusing on the perioperative, surgical, and long-term management of patients with long-gap esophageal atresia. In line with the first ERNICA consensus conference,[14,15] this conference was based on two keystones: (1) on evidence from literature and (2) on expert opinion.

Participants of this conference achieved general consensus (defined by $\geq$ 75% of votes scoring 6–9) in 93% of all items ($n = 97$) which indicates a considerable level of agreement and suggests predominantly homogeneous approaches in ERNICA institutions. This is supported by the high rate of total agreement (defined as 100% consensus among voters) of 58% of all items. A maximum range of voting from 1 to 9, indicating widely diverse opinions, was evident for only 20% of items which is low considering the huge variability of current treatment strategies.

Consensus was not reached in only 7/97 votes (7%), and all of these statements were discussed controversially (range 1–9; except one with a range 2–9):

Table 9 Priorities for further research

	Domain	Topic
1	Diagnostics	Optimal approach for gap measurement
2	Definitions	Comprehensive definition of "long-gap esophageal atresia"
3	Esophageal reconstruction - *Initial management before reconstruction*	Counseling of parents (ideally including the involvement of patient support groups)
4	Esophageal reconstruction - *Delayed primary anastomosis*	Evidence for routine insertion of a transanastomotic tube
5	Esophageal reconstruction - *Delayed primary anastomosis*	Evidence for routine placement of a chest drain
6	Esophageal reconstruction - *Lengthening techniques*	Early and long-term outcome of different esophageal lengthening techniques
7	Esophageal replacement	Evidence for optimal surgical technique for esophageal replacement
8	Esophageal replacement - *Gastric transposition*	Early and long-term outcome after gastric tube formation as an option for esophageal replacement
9	Esophageal replacement - *Gastric transposition*	Evidence for insertion of a transanastomotic tube during gastric transposition
10	Esophageal replacement - *Gastric transposition*	Relevance of pyloroplasty (Mikulicz) during gastric transposition
11	Postoperative management	Evidence for routine postoperative contrast study of the esophagus before initiation of oral feeding
12	Postoperative management	Timing of the initiation of oral feeding
13	Follow-up	Duration of postoperative antacid therapy
14	Follow-up	Mode of tapering the postoperative antacid therapy
15	Follow-up	Evidence for peri-interventional antibiotic prophylaxis in balloon or semirigid dilatation for anastomotic stricture
16	Follow-up	Application of indwelling balloon dilatation, endoscopic knife, and surgical resection and reanastomosis in cases of recurrent anastomotic stricture

Gap Measurement

Accurate measurement of gap length is critical for operative planning for long-gap esophageal atresia.[29,30] It is still a matter of debate whether preoperative contrast study to evaluate the upper esophageal pouch is needed. McDuffie et al stressed the high risk of aspiration[31] which can be disastrous, as exemplified in a case-reported death of an infant.[32] Moreover, Gross et al postulated that gap measurement with contrast media is not reliable as the lower pouch might be underestimated in the absence of reflux.[29] Based on these arguments, the need for contrast studies for diagnostics has been essentially eliminated,[33] and hence, gap measurement by contrast study of the upper and lower pouches was not considered to be a viable option by the participants of the conference.

Definition of "Long-Gap" Esophageal Atresia

Efforts have been made to define long-gap esophageal atresia as precisely as possible and not to accept subjective terms such as "inability to achieve primary end-to-end anastomosis."[3,4,6,9,34–36] However, there is no universally accepted methodology for determining either the gap length or what constitutes long-gap esophageal atresia.[3] The APSA Committee—based on its members' opinion—stated that the nomenclature "*Long Gap Esophageal Atresia*" should not be reserved for Gross type A atresia exclusively, which is in line with the results of the participants' votes. The INoEA working group recently reaffirmed the unclear definition and suggested to define any esophageal atresia that has no intra-abdominal air should be considered a long gap.[5]

This statement reached complete consensus by all participants. However, it was additionally stated that this definition should not confine long-gap esophageal atresia to pure atresia (Gross type A or B), but should allow inclusion of esophageal atresia with tracheoesophageal fistula with a wide gap, respectively, with different anatomic configurations.[1,36–39] Participants decided not to vote on this proposed term of "wide gap" to avoid more confusion. As a result of a highly controversial discussion on the definition of long-gap esophageal atresia, consensus was reached on important statements. It must be emphasized, however, that the condition of long-gap esophageal atresia is not fully depicted yet by these definitions.

Esophageal Reconstruction

There is no consensus on the preferred method of esophageal reconstruction to date.[2,4,8,26,40–42] No randomized controlled trials comparing different approaches of surgical repair or comparing the various techniques used to manage long-gap esophageal atresia are available.[8] Fundamentally, it must be

stated that preservation of the native esophagus should be aimed before considering any replacement technique[9,28,43] as "*no other conduit can replace its function in transporting food from the oral cavity to the stomach satisfactorily*".[44] In this light, it is interesting to mention that thoracoscopic pouch mobilization and placement of traction sutures are considered a novel technique that shows promise, but should only be performed in specialized centers with prospective review and reporting of outcomes (►Table 3, f).

Various different techniques deal with esophageal replacement, reflecting that none has been suggested to be ideal, and the patients are left with many challenges to overcome.[2,9,43,45,46] In this context, participants voted on the preferred option for esophageal replacement (►Fig. 1). The majority of participants voted for gastric transposition (56%), followed by jejunal interposition (25%) and finally followed by colonic interposition (19%). The voting result corresponds to the current literature as multiple studies confirmed that the stomach is the preferred organ for esophageal replacement.[8,47–50] In addition, the high rate of abstention from voting (30%) indicates the disagreement of preferred surgical options. The latest systematic review of the surgical treatment of long-gap esophageal atresia by Stadil et al aimed to compare the postoperative complications related to the different methods within the first postoperative year.[40] Fifty-seven articles were included involving 326 patients with Gross types A and B long-gap esophageal atresia.[40] Delayed primary anastomosis was the most applied surgical method (68.4%) in both types, followed by gastric transposition (8.3%) which is in line with the statements achieved during the consensus conference.

Literature provides some evidence for colonic interposition to be a surgical option comparable to other replacement techniques.[2,51,52] Nonetheless, the INoEA working group stated that colonic interposition is mainly reserved as a last option, when all other techniques have failed or are considered unfeasible.[5] After extensive discussion, participants of this conference did not reach consensus on colonic interposition as a viable option for esophageal replacement mainly driven by evidence from literature[5,53–55] and individual experience.

Placement of Chest Drain

Routine placement of a chest drain in delayed primary anastomosis was also controversially discussed. Several studies postulated that a chest drain does not alter early postoperative complications after repair of esophageal atresia with tracheoesophageal fistula.[56–58] Participants generally agreed that the placement of a chest drain for delayed primary anastomosis has to be valued differently compared with primary repair due to higher risk of anastomotic leakage based on an anastomosis under tension. Even though a clear majority of voters supported the routine placement of a chest drain, consensus was not reached on this item.

Postoperative Contrast Study

Evidence on "routine postoperative contrast study of the esophagus before the initiation of oral feeding" is scarce.[59,60] Yanchar et al presented a study including 90 patients who underwent esophageal atresia repair and postoperative upper gastrointestinal contrast study before consideration of oral feeding.[60] They concluded that the use of early routine contrast studies, with no suspicion of a problem, has little value in terms of predicting complications or future clinical course.[60] These findings support the participants' votes not advocating for a routine postoperative contrast study in the absence of any clinical findings.

Given the complexity of this patient population with significant morbidity and associated anomalies, treatment and long-term follow-up should be managed by specialized and multidisciplinary teams.[9,11,28,61] There are growing demands for referral of esophageal atresia patients to designated centers of expertise just as it is common practice in the Netherlands or in France, and these demands are even stronger for patients with long gap.[3,4,8,9,14,15,62,63] These repeatedly expressed requirements to fulfill the criteria of optimal patient care are reflected in the consensus statements in the domain *Framework*.

Although the consensus meeting was focused entirely on long-gap congenital esophageal atresia, esophageal reconstruction is also often necessary for children with caustic injury, button battery ingestion, or other acquired/iatrogenic esophageal damage. Although the statements generated are not directly applicable to such children, some of the statements might be considered as relevant and useful in the absence of any specific consensus guidelines on treatment of children with acquired esophageal damage.

As stated previously, the strength of this conference is the pool of participating specialists with extensive expertise in this field.[14,15] The multidisciplinary approach was highly valued in the first ERNICA consensus conference allowing discussion from various perspectives. Considering this beneficial methodological approach, the group of participants had been extended to representatives of the specialty neonatology and pediatric pulmonology. Further advantages, such as the methodology (characterized by meticulous item generation and prioritization, systematic literature search, and anonymous voting), ability to modify wording of statements via the online voting system, abstention from voting in case of lacking expertise, and involvement of representatives of the patient support groups ensure the high quality of results and indicate the great validity of votes.

Nonetheless, it has to be emphasized that the results of the conference were mainly based on expert opinion, and not on evidence.[14,15]

Conclusion

Evidence supporting best practices for long-gap esophageal atresia is weak. We hereby present consensus statements on the perioperative, surgical, and long-term management of patients with long-gap esophageal atresia based on a critical evaluation of the current literature. Areas of controversy were identified for future research. Substantial statements regarding esophageal reconstruction or replacement and follow-up were formulated which may contribute to optimized and uniform patient care.

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Conflict of Interest

None declared.

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