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ERNICA Consensus Conference on the Management of Patients with Esophageal Atresia and Tracheoesophageal Fistula: Diagnostics, Preoperative, Operative, and Postoperative Management

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Abstract

Introduction Many aspects of the management of esophageal atresia (EA) and tracheoesophageal fistula (TEF) are controversial and the evidence for decision making is limited. Members of the European Reference Network for Rare Inherited Congenital Anomalies (ERNICA) conducted a consensus conference on the surgical management of EA/TEF based on expert opinions referring to the latest literature.

Materials and Methods Nineteen ERNICA representatives from nine European countries participated in the conference. The conference was prepared by item generation, item prioritization by online survey, formulation of a final list containing the domains diagnostics, preoperative, operative, and postoperative management, and literature review. The 2-day conference was held in Berlin in October 2018. Anonymous voting was conducted via an internet-based system. Consensus was defined when 75% of the votes scored 6 to 9.

Keywords

- esophageal atresia
- European Reference Network on Rare Inherited and Congenital Anomalies (ERNICA)
- surgical management
- evidence
- consensus conference

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Results Fifty-two items were generated with 116 relevant articles of which five studies (4.3%) were assigned as level-1evidence. Complete consensus (100%) was achieved on 20 items (38%), such as TEF closure by transfixing suture, esophageal anastomosis by interrupted sutures, and initiation of feeding 24 hours postoperatively. Consensus $\geq$75% was achieved on 37 items (71%), such as routine insertion of transanastomotic tube or maximum duration of thoracoscopy of 3 hours. Thirteen items (25%) were controversial (range of scores, 1–9). Eight of these (62%) did not reach consensus.

Conclusion Participants of the conference reached significant consensus on the management of patients with EA/TEF. The consensus may facilitate standardization and development of generally accepted guidelines. The conference methodology may serve as a blueprint for further conferences on the management of congenital malformations in pediatric surgery.

Introduction

Esophageal atresia is a rare congenital condition with an estimated prevalence varying between 1 and 2 in 5,000 live births in Europe.[1,2] The malformation is characterized by a discontinuity of the esophagus with or without tracheoesophageal fistula. Since the first successful primary repair of esophageal atresia in 1941, improvements in operative and perioperative care have led to better outcomes, and mortality has decreased to a single-digit range[3–5] with most deaths due to comorbidities. Evolution from concerns about mortality to concerns about morbidity and quality-of-life issues has occurred,[5] and long-term morbidity remains high until adulthood.[5,6] Morbidities include esophageal,[7] gastrointestinal,[8] pulmonary,[9,10] and various developmental[11] problems which may have a considerable impact on the quality of life of patients and their families.[6,12–15] Therefore, esophageal atresia is no longer a mere neonatal surgical problem but rather lifelong requires attention in individual patients.[5,16]

Well-designed clinical trials dealing with diagnostic and therapeutic concepts for patients with esophageal atresia are still scarce[3,17–19] and generally accepted algorithms are lacking. As a result, there is a variety of coexisting protocols on the perioperative and surgical management of patients with esophageal atresia based on opinion rather than on evidence.[20] Diversity of concepts and the use of different operative techniques even for the same type of atresia have a significant impact on the course of the disease in single patients.

The European Reference Network on Rare Inherited and 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.[21] ERNICA is one of 24 European Reference Networks cofunded by the European Union (Health Program), and involves teams from 20 European hospitals from 10 member states.[21] The mission is to promote optimal patient care for rare inherited and congenital digestive track-related disorders from pediatric age to adulthood providing high-quality and accessible education, supporting research, improving clinical standards, and services and reducing health inequalities in Europe.[22–25]

The ERNICA Workstream, *Congenital Malformations and Diseases of the Esophagus*, is mainly focused on patients with esophageal atresia. During recent ERNICA meetings in Rotterdam (April 2017), Helsinki (November 2017), and Stockholm (April 2018), it became evident that protocols dealing with the management of esophageal atresia considerably differ even between ERNICA institutions. Therefore, the members of ERNICA agreed to establish consensus on all relevant aspects of the surgical management of patients with esophageal atresia as an urgent objective of ERNICA.

The aim of this first ERNICA consensus conference was to establish consensus on the relevant aspects regarding the treatment of children with esophageal atresia and tracheoesophageal fistula based on expert opinions referring to the latest evidence from current literature. The conference was systematically prepared to include clinical and nonclinical members of ERNICA. In particular, representatives of several national patient support groups, which have become full members of ERNICA,[26,27] have been invited to participate in the conference.

Materials and Methods

Participants

Members of the ERNICA Workstream, *Congenital Malformations and Diseases of the Esophagus*, were invited to take part in the preparation and implementation of the conference. In total, 19 participants originating from 15 institutions and nine countries confirmed participation. Three representatives from national patient support groups acting under the umbrella of the Federation of Esophageal Atresia and Tracheo-Esophageal Fistula Support Groups (EAT),[26] were included. Altogether 14 pediatric surgeons, one pediatric gastroenterologist, three representatives of patient support groups, and one nonsurgeon pediatric surgery academic took part in all steps of the preparation and the conference itself. The conference took place in Berlin on October 25 and 26, 2018.

The preparation and implementation of the conference included three steps as follows: (1) generation of a list of items; (2) prioritization of the items; (3) discussion of all items during the conference and formulation of statements; and (4) anonymous voting.

Focus of the Conference

The conference dealt exclusively with the management of patients with esophageal atresia with tracheoesophageal fistula who undergo primary anastomosis. Items dealt with the surgical, perioperative, and long-term management of these patients, and the conference was focused on operative and perioperative aspects. The conference did not deal with the management of other forms of esophageal atresia, such as long-gap atresia or those with secondary anastomosis.

Item Generation

A systematic literature search was performed by C.D. and B.U. to identify relevant aspects in the management of esophageal atresia with tracheoesophageal fistula. A PubMed literature search was conducted for the years 2008 to 2018 using the keywords "esophageal/oesophageal atresia," "tracheoesophageal fistula," "diagnostics," "management," "surgery," "operation," "complications," "outcome," "experience," and "follow-up/follow up." In case of absent relevant literature, the time period was extended to include more historic literature. Reports on long-gap esophageal atresia only, non-English articles, case reports, and reviews without original patient data were excluded.

After literature search, a preliminary item list was developed which was presented during an ERNICA conference in Stockholm on the April 18 to 20, 2018. Members of the Workstream *Congenital Malformations and Diseases of the Esophagus* discussed this list in detail. The list was modified according to this discussion by exclusion of some suggested items and inclusion of new items which were considered to be relevant. The list was distributed to all participants who were invited to revise the items and to test for plausibility.

Finally, items were attributed to the following domains: (1) diagnostics, (2) preoperative management after confirmation of diagnosis, (3) operative management, (4) postoperative management, (5) follow-up; and (6) miscellaneous. The consensus results and discussion for the domains diagnostics, preoperative, operative, and postoperative management are presented in this manuscript; the domains follow-up and miscellaneous will be addressed in a separate manuscript as the methodological approaches differed.

Item Prioritization

The online REDCap electronic data capture tools[28] hosted at University College London was used for prioritization of the items. Priority was assigned to each item by all participants using a five-point Likert's scale (1 as the highest priority and 5 as the lowest priority). Scoring each item was compulsory. Thereafter, the final list of items to be discussed at the conference was set up by C.D., B.M.U., and S.E., taking the prioritization into account. The threshold for exclusion or inclusion of items was scoring as "lowest" or "highest" priority, respectively, by two or more participants; entirely new items were added after suggestion by two or more participants independently.

Identification of Relevant Literature for Item Discussion

Prior to the conference, two participants were allocated to each domain (domain leaders). Domain leaders performed a literature research and identified the most relevant publications for each item of their domain. Publications with the highest grade of evidence according to the CEBM (Center for Evidence-Based Medicine) classification were suggested to be preferred. The highest grade of evidence was defined as level-1 evidence derived from studies with a "high" quality of evidence ("further research is unlikely to change our confidence in the estimate of effect"). Level-1 evidence was derived from systematic reviews (with homogeneity) or meta-analyses of randomized controlled trials (CEBM level-1a), a well-designed individual randomized controlled trial (with narrow confidence interval; CEBM level-1b), or all or nonrandomized controlled trials (CEBM level-1c).[29]

Literature was pooled by C.D. After sorting, it was distributed and made available to all participants via a DropBox link prior to the conference.

Conference, Voting, and Consensus

The final list of domains, items, and references was accessible to all participants via an online link prior to and throughout the conference. Each domain was discussed in a separate session. Domain leaders introduced the literature and relevant aspects on single items. Subsequently, participants discussed each item and formulated a statement to facilitate voting. The wording of the statements evolved during group discussion.

Participants voted on each statement via the internet-based system VoxVote.[30] An event code was provided and participants were able to vote using either an internet browser, or dedicated apps for Android or iOS platforms. The wordings of the statements on items were updated during the discussion by the nonsurgical academic (S.E.) who did not vote. Participants were able to vote using a 1 to 9 scale (9 meaning "fully agree" and 1 "strongly disagree"). Participants were suggested not to vote on an individual item when they felt that it was not an item that they had expertise or an opinion on. Therefore, participants were allowed to vote online for "no relevant expertise on this statement." As a result, the number of scoring participants varied for single statements.

Consensus was defined as $\geq$ 75% of those voting having scored 6, 7, 8, or 9, excluding those who declared no relevant expertise on that statement.

The voting was anonymous and scores were not visible to the participants during the voting process. The final scores and the consensus results were shown to all participants after all votes were obtained, but individual scores remained anonymous. The 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 ERNICA conference in Stockholm on the April 18 to 20, 2018 resulted in a total of 41 items.

After the online prioritization phase, two items were excluded. Following the participants' suggestions, seven items were added as new items. Consequently, the list included 46 items prior to the conference for which literature was obtained and circulated.

As a result of the presentations by the domain leaders and discussion during the conference, five items were excluded, and 11 items were added as new items as some items were split into several separate questions. Finally, 52 items were confirmed for voting and included six items in the domain diagnostics, eight items in the domain preoperative management, 28 items in the domain operative management, and 10 items in the domain postoperative management (►Tables 1–4).

Relevant Literature

A total of 116 relevant manuscripts were selected by literature search conducted by all participants (**Supplementary Material**, available in the online version). Ten studies among them had been identified for several items, and 10 were quoted for more than one domain. In summary, 18 articles addressed the domain diagnostics, 30 articles for preoperative management, 63 articles for operative management, and 17 articles for postoperative management.

CEBM level-1 evidence was only available for four items (7.7%) and included four single randomized controlled trials and one pilot randomized trial which represented 4.3% of all selected articles (►Table 5). Ten level-2 evidence studies (8.6%), 26 level-3 evidence studies (22.4%), 43 level-4 evidence studies (37.1%), and 11 level-5 evidence studies (9.5%) were also included. Moreover, 15 review articles (12.9%) and six book chapters (5.2%) have been considered to be relevant for the discussion, despite not including original data (►Fig. 1).

Consensus

Total agreement, defined as 100% consensus amongst voters, was achieved on 20 items (38%) and consensus (≥ 75% of those voting having scored 6–9) on 37 items (71%). Thirteen items (25%) were particularly controversial in that the votes ranged from 1 to 9; in eight of these no consensus was reached. Detailed results are summarized in ►Tables 1–4.

Abstention

Participants were supposed not to vote when they felt a lack of competence. One or more participants declared "no relevant expertise on this statement" on the online voting system on 48 (92.3%) questions—for three (5.8%) questions, one participant abstained; for two (3.8%) questions, two participants abstained; in 21 questions (40.4%) three participants abstained; in 20 (38.5%) processes, four participants abstained; in two questions (3.8%), five participants abstained from voting.

Controversial Items Discussed without Voting

Several items were discussed that were controversial and the participants agreed verbally that more data from future research would have to be available before a meaningful question and vote could be conducted (►Table 6). The discussion included in particular the following aspects:

Diagnostics

The participants agreed that magnetic resonance imaging represents an important tool in identifying anomalies of the aortic arch and its branches, and might be considered as the imaging technique of choice when planning surgical management, especially in cases of associated cardiac anomalies. However, as the evidence from the literature[31] and personal experience on the routine use of magnetic resonance imaging in the preoperative workup of esophageal atresia patients was limited; it was decided not to vote on this item.

Table 1 Diagnostic procedures in patients with suspected esophageal atresia

	Diagnostics	Consensus	%	Votes	Median (range)
1	A nasogastric tube 10 Fr 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 to exclude 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)

Abbreviation: EA, esophageal atresia.

Table 2 Preoperative management of patients with esophageal atresia after confirmation of the diagnosis before transfer to the operation theater

	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 favored	+	100	15/15	9 (9–9)
10	If assisted ventilation is required, preference should be given to intubation rather than to noninvasive 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)

Table 3 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 thoracotomy	+	100	15/15	9 (8–9)
22	Entry through the 4th 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 tracheoesophageal fistula should preferably be closed by transfixing suture	+	100	14/14	9 (6–9)
27	The esophageal anastomosis should be preferably performed with absorbable sutures	+	85.7	12/14	8 (1–9)
28	The esophageal anastomosis should be preferably performed with interrupted sutures	+	100	14/14	9 (6–9)
29	A transanastomotic tube should be routinely inserted	+	80	12/15	8 (1–9)
30	A chest drain should be routinely placed	–	21.4	3/14	1 (1–9)
31	The thoracoscopic approach is a viable option	+	87.5	14/16	9 (5–9)
32	The thoracoscopic approach should be only performed if suitable expertise is available	+	100	17/17	9 (6–9)

Table 3 (*Continued*)

	Operative management	Consensus	%	Votes	Median (range)
33	The thoracoscopic approach offers the advantage of magnification compared with the conventional approach	+	92.9	13/14	9 (5–9)
34	The thoracoscopic approach offers the advantage of faster recovery compared with the conventional approach	–	53.3	8/15	6 (1–9)
35	The thoracoscopic approach offers the advantage of better cosmesis compared with the conventional approach	+	94.1	16/17	9 (5–9)
36	The thoracoscopic approach offers the advantage of less musculoskeletal sequelae compared with the conventional approach	+	86.7	13/15	8 (5–9)
37	The maximum insufflation pressure of CO_2 during thoracoscopy should not exceed 5 mm Hg	+	100	14/14	9 (6–9)
38	Maximum duration of thoracoscopic operation should be 3 h	+	92.9	13/14	8 (4–9)
39	The thoracoscopic approach has the disadvantage of longer operative time compared with the conventional approach	+	92.9	13/14	7.5 (2–9)
40	The thoracoscopic approach has a negative pathophysiological impact (acidosis, cerebral oxygenation impairment) compared with the conventional approach	–	30.8	4/13	5 (1–7)
41	The thoracoscopic approach has the disadvantage of a higher complication rate compared with the conventional approach	–	35.7	5/14	3.5 (1–8)
42	There is no place for routine fundoplication in patients with EA during the initial operation.	+	100	18/18	9 (8–9)

Abbreviation: EA, esophageal atresia.

Table 4 Postoperative management of patients with esophageal atresia

	Postoperative management	Consensus	%	Votes	Median (range)
43	Postoperative ventilation and relaxation should not be routine and should be 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 refistula, or missed upper pouch fistula, if suspected	+	93.8	15/16	9 (3–9)
51	A refistula 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)

Table 5 Literature meeting the criteria of CEBM level 1 evidence[a]

Statement	Domain	Reference	Study type
The muscle-sparing approach is the recommended approach for conventional thoracotomy	Operative management	Askarpour et al[47]	RCT
The azygos vein should be preserved whenever possible	Operative management	Upadhyaya et al[48] Sharma et al[49]	RCT RCT
The thoracoscopic approach has the disadvantage of a negative pathophysiological impact (acidosis, cerebral oxygenation impairment) compared with the conventional open approach	Operative management	Bishay et al[61]	Pilot RCT
An anastomotic leakage within the first 4 postoperative days may be considered for surgical revision	Postoperative management	Vaghela et al[36]	RCT

Abbreviations: CEBM, center for evidence-based medicine; RCT, randomized controlled trial.
[a]In accordance with the Oxford CEBM levels of evidence as published in 2009.[29]

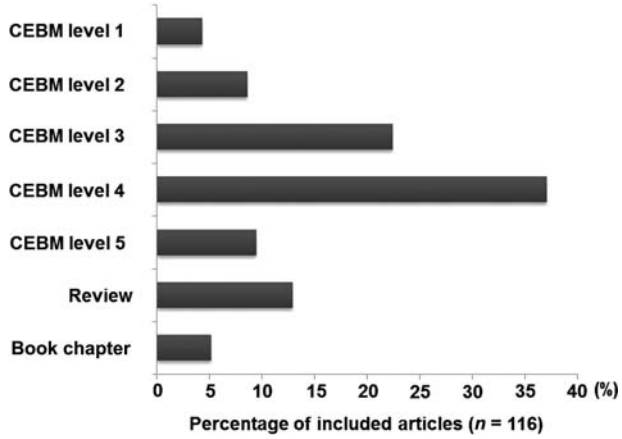

Fig. 1 CEBM level of evidence of articles considered as relevant for the consensus statements. CEBM, Center for Evidence-Based Medicine. In accordance with the Oxford CEBM levels of evidence as published in 2009.[29]

Preoperative Management

Several participants postulated that preoperative measurement of the esophageal gap length may promote comparability between centers, may reduce the incidence of unsuspected difficult cases, and may contribute to a general definition of long-gap esophageal atresia.[32,33] After extensive discussion, it was decided not to vote on a statement on preoperative gap measurement as a routine procedure due to the lack of data and literature on this item.

Operative Management

A vote on routine posterior tracheopexy during primary repair of esophageal atresia was rejected. Most participants felt that the available data were scarce,[34,35] and the experience with this new technique remained too limited to allow a meaningful vote.

Postoperative Management

The management of anastomotic leakage was discussed extensively. No consensus was achieved on surgical revision of anastomotic leakage within the first 4 postoperative days. Vaghela et al[36] presented data from a randomized controlled trial on the application of glycopyrrolate as a therapeutic option for patients with postoperative anastomotic leakage. However, despite this evidence, the conference participants felt that is too early to vote on the role of glycopyrrolate.

Participants supported the concept that a clinical checklist of tasks and examinations should be compiled before initial hospital discharge, including abdominal and renal ultrasound, and resuscitation training for caregivers (►**Table 4**). The full content of such a list remained a matter of discussion and agreement could not be achieved.

Discussion

According to the Council of Europe a medical consensus is a public statement on a particular aspect of medical knowledge that is generally agreed upon as an evidence-based, state-of-the-art knowledge by a representative group of experts in that area.[37] Its main objective is to counsel physicians on the best possible and acceptable way to address a particular decision-making area for diagnosis, management, or treatment.[38] Most recently, multiple consensus conferences have been organized both in the field of adult[39,40] and pediatric medicine.[41,42]

Table 6 Controversial discussion items leading to exclusion from voting due to inability to formulate a meaningful voting question

Domain	Discussed item	Result
Diagnostics	Role of routine preoperative magnetic resonance imaging	No voting
Preoperative management	Measurement of the gap length before operation	No voting
Operative management	Routine posterior tracheopexy during primary repair of esophageal atresia	No voting
Postoperative management	Application of glycopyrrolate as a therapeutic option for anastomotic leakage	No voting
	Content of a checklist for first discharge	No voting

Importantly for esophageal atresia, Krishnan et al published in 2016 the ESPGHAN–NASPGHAN guidelines for the evaluation and treatment of gastrointestinal and nutritional complications in children with esophageal atresia and tracheoesophageal fistula.[18] The guidelines were set up during two consensus meetings using the nominal voting technique. Expert opinion was used where no randomized controlled trials were available to support the recommendations.[18] This consensus statement focused on the medical aspects of follow-up, and few surgical aspects were included. We hereby present for the first time the results of a consensus conference focused on surgical aspects of the management of patients with esophageal atresia with tracheoesophageal fistula. In line with the ESPGHAN–NASPGHAN guidelines, this conference was based on the two following keystones: (1) on expert opinion and (2) on evidence from literature.

Modern medicine increasingly places emphasis on evidence-based medicine,[43] defined by Sackett et al as "*(…) the conscientious, explicit and judicious use of current best evidence in making decisions about the care of individual patients*."[44] Nonetheless, the paucity of high-level evidence in the literature on pediatric surgical procedures was highlighted in 1999, when Hardin et al[45] reported that only 0.3% of the literature relevant for pediatric surgery consisted of CEBM level-1 evidence studies. In 2010, Ostlie and St. Peter[46] demonstrated that prospective articles represented less than 0.05% of all pediatric surgical literature. In line with these findings, literature meeting the criteria of CEBM level-1 evidence was only available for four out of 52 items (7.7%) that were discussed at this conference. Consequently, the votes of the participants of the conference were based on a combination of lower level evidence (e.g., retrospective reviews) combined with expert opinion.

The voting resulted in 100% consensus for 20 (38%) of items. This indicates a considerable level of agreement but also a certain diversity of opinions among specialists in the field of esophageal atresia. However, general consensus (defined by $\geq$ 75% of votes scoring higher 6–9) was achieved for 71% of items suggesting predominantly homogeneous approaches in ERNICA institutions. A maximum range of voting from 1 to 9, indicating widely diverse opinions, was evident for only 25% of cases.

Interestingly, consensus was achieved on most, but not on all items with level-1 evidence in the literature. Askarpour et al recently confirmed better outcomes of muscle-sparing versus standard posterolateral thoracotomy in newborns.[47] This statement was adopted unanimously by the participants of the conference. However, advantages of the preservation of the azygos during esophageal atresia repair were confirmed in two randomized trials[48,49] but no consensus could be achieved on this topic. Several participants questioned the quality of both trials and as a result consensus was not achieved.

Dingemann and Ure[50] reported on low CEBM levels of evidence in the field of endoscopic pediatric surgery. Dingemann et al[20] also demonstrated that only retrospective comparative studies pursuant to CEBM level 3 were available on pediatric video-assisted thoracoscopic surgery. With regard to the management of esophageal atresia, it has been stated that the best available evidence comparing the minimally invasive versus the open approach for esophageal atresia repair was CEBM level 3.[51] Since then, minimally invasive esophageal atresia repair has been subject of numerous studies.[52–60] Despite the low level of evidence of these reports, consensus was achieved on several advantages of thoracoscopic esophageal atresia repair based on the experience of the participants. One pilot randomized controlled trial on 20 neonates dealt with potential disadvantages of the technique.[61] Patients with congenital diaphragmatic hernia but not those with esophageal atresia had severe intraoperative and prolonged hypercapnia and acidosis. However, it should be emphasized that pulmonary compromised patients had been included in this study, as well as the intraoperatively applied insufflation pressures were rather high which could explain the described poor outcome. Taking this into account, the participants of the conference also felt that the numbers of patients of this pilot trial were too small to draw valid conclusions on management of infants with esophageal atresia.

The management of anastomotic leakage is still the subject of ongoing discussions.[62,63] Vaghela et al[36] recommended the application of glycopyrrolate as a therapeutic option. Despite these results, the conference participants felt that is too early to vote on the role of glycopyrrolate.

In 2014, the EUPSA international survey on the management of esophageal atresia demonstrated that approximately 60% of respondents measure the gap intra-, but not preoperatively.[17] After a controversial discussion, it was decided not to vote on a statement on preoperative gap measurement as a routine procedure.

The issue of a routinely performed posterior tracheopexy during primary repair of esophageal atresia was a matter of debate. Shieh et al concluded from their experience with 118 patients that posterior tracheopexy should be selectively considered at the time of initial repair.[34,39] Tytgat et al recently presented nine patients who underwent thoracoscopic posterior tracheopexy during primary esophageal atresia repair.[35] The authors postulated that this technique prevents potential sequelae of mild to severe tracheomalacia and a second surgical procedure. The participants felt that evidence was too low to allow voting and that potential disadvantages of the technique remain unclear.

A clinical checklist may serve as an aid for clinicians not to miss essential measures before discharge. Recent literature provides an example for such similar standardized checklist.[64] Conference participants strongly supported the concept of a checklist of tasks and examinations to be used before the first discharge. However, consensus could not be achieved on which items should be included, except for abdominal and renal ultrasounds and resuscitation training for caregivers.

The strength of this conference is the pool of participating specialists with extensive expertise in the field. The multidisciplinary approach allowed discussing items from various perspectives. An additional advantage of this conference was the methodology characterized by meticulous item generation and prioritization, systematic literature search and anonymous voting ensuring a high quality of results. The ability

to modify wording of questions flexibly and rapidly via the online voting system was an advantage, which allowed consensus to be reached on 37 items, with a further 15 voted on without consensus. The approach that we used might be advocated for other surgical consensus meetings, in which there might be a somewhat higher number of (often technical) items than for medical consensus meetings in which consensus statements might be generated on, for example, a treatment approach. Abstention from voting in case of lacking expertise additionally indicates a high validity of votes. It may be speculated that representatives of the patient support groups and the pediatric gastroenterologist refrained from voting in cases of entirely surgical issues. Finally, representatives of patient support groups were able to contribute to the discussions and voting process from the patients' perspective. The items on which either no consensus was reached, or where it was felt that there was insufficient evidence to hold a meaningful vote, could be suggested to be future areas which are priorities for future research. There are several drawbacks of the conference. First, as the conference was focused on pediatric surgeons, only one pediatric gastroenterologist and no pediatric pulmonologist, pediatric anesthesiologist, and neonatologist were involved. Numerous perioperative aspects may have been discussed differently by representatives from nonoperative fields. Second, the results of the conference were mainly based on expert opinion. Therefore, the statements of conference participants must be considered as opinions of ERNICA representatives only.

Conclusion

Participants of this ERNICA conference reached significant consensus on the pre-, peri-, and postoperative management of patients with esophageal atresia and tracheoesophageal fistula who undergo primary anastomosis. The consensus may facilitate standardization and development of generally accepted guidelines for patient care. The conference methodology may serve as a blueprint for further conferences on the management of other congenital malformations in pediatric surgery.

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Conflict of Interest

None declared.

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