European Reference Network
for rare or low prevalence complex diseases

Network Inherited and Congenital Anomalies (ERNICA)

ERNICA CONSENSUS CONFERENCE ON THE SURGICAL MANAGEMENT OF PATIENTS WITH ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA

WITH EXPLANATIONS FOR PATIENTS AND FAMILIES

Version 1. 05-10-2022

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. Tracheoesophageal fistula is when a child is born with an abnormal connection between the trachea (also known as the 'windpipe') and the esophagus. Esophageal Atresia and tracheoesophageal fistula are areas of focus for the ERNICA network. Most children born with tracheoesophageal fistula also have Esophageal Atresia.

In 2018, a conference took place which was attended by 19 representatives from ERNICA institutions (across 9 European countries) who have extensive experience in the treatment of patients with esophageal atresia. This group of participants included pediatric surgeons, a pediatric gastroenterologist and representatives of patient support groups. The aim of this conference was to establish consensus on the surgical management of patients with Esophageal Atresia and tracheoesophageal fistula, by reviewing the latest scientific evidence and considering expert opinion.

An anonymous online voting process took place and consensus was reached on 60 aspects related to the care of patients for Esophageal Atresia and tracheoesophageal fistula. Two scientific papers which report the findings were published in The European Journal of Pediatric Surgery in 2019.

1. *ERNICA Consensus Conference on the Management of Patients with Esophageal Atresia and Tracheoesophageal Fistula: Diagnostics, Preoperative, Operative, and Postoperative Management* (published online July, 2019): HTML version , PDF.
2. *ERNICA Consensus Conference on the Management of Patients with Esophageal Atresia and Tracheoesophageal Fistula: Follow-up and Framework* (published online November, 2019): HTML version , PDF

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

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). 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 the KEKS website: https://www.keks.org/keks-hilft/projekte/ernica-leitlinien/*

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DIAGNOSIS AND MANAGEMENT BEFORE, DURING AND AFTER SURGERY

DIAGNOSIS

	STATEMENTS	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 EA.	+	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 AS A PREOPERATIVE DIAGNOSTIC PROCEDURE.	+	
3	AN ULTRASOUND OF THE ABDOMEN (INCLUDING KIDNEY/URINARY TRACT) SHOULD BE ROUTINELY PERFORMED AS A PREOPERATIVE DIAGNOSTIC PROCEDURE.	-	AN ULTRASOUND OF THE KIDNEYS AND OTHER ABDOMINAL ORGANS DOES NOT HAVE TO BE DONE BEFORE THE OPERATION. AN ULTRASOUND IS A WAY OF TAKING IMAGES INSIDE THE BODY.
4	AN ULTRASOUND OF THE SPINE SHOULD BE ROUTINELY PERFORMED AS A PREOPERATIVE DIAGNOSTIC PROCEDURE.	-	AN ULTRASOUND OF THE SPINE DOES NOT HAVE TO BE DONE BEFORE THE OPERATION.
5	ECHOCARDIOGRAPHY SHOULD BE ROUTINELY PERFORMED AS A PREOPERATIVE DIAGNOSTIC PROCEDURE, 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.
6	A CONTRAST STUDY OF THE 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.

PRE-OPERATIVE MANAGEMENT*

*AFTER THE PATIENT HAS BEEN DIAGNOSED WITH EA AND BEFORE THE PATIENT IS TRANSFERRED TO THE OPERATING ROOM FOR SURGERY

	STATEMENTS	CONSENSUS/NO CONSENSUS	EXPLANATIONS FOR PATIENTS/ PARENTS
7	A REPLOGLE TUBE SHOULD BE ROUTINELY PLACED INTO THE UPPER ESOPHAGEAL POUCH TO ALLOW CONTINIOUS 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.
8	PREOPERATIVE ANTIBIOTIC PROPHYLAXIS SHOULD BE ROUTINELY ADMINISTERED AS SOON AS THE DIAGNOSIS IS ESTABLISHED.	-	
9	SPONTANEOUS BREATHING SHOULD ROUTINELY BE FAVOURED.	+	
10	IF ASSISTED VENTILATION IS REQUIRED, PREFERENCE SHOULD BE GIVEN TO INTUBATION RATHER THAN TO NON-INVASIVE VENTILATION.	+	
11	TRACHEOBRONCHOSCOPY UNDER SPONTANEOUS BREATHING SHOULD BE PERFORMED PREOPERATIVELY TO EVALUATE TRACHEOMALACIA.	-	ASSESSMENT OF THE AIRWAY FOR FLOPPINESS IS **NOT** NECESSARY AS A FIRST PROCEDURE.
12	A CENTRAL VENOUS LINE SHOULD BE ROUTINELY PLACED PREOPERATIVELY.	-	
13	AN ARTERIAL LINE SHOULD BE ROUTINELY PLACED PREOPERATIVELY.	-	
14	DURING PREOPERATIVE COUNSELLING PARENTS SHOULD BE ROUTINELY INFORMED ABOUT DIFFERENT SURGICAL OPTIONS, SUCH AS OPEN AND THORACOSCOPIC REPAIR.	+	ALL POSSIBLE SURGICAL PROCEDURES SHOULD BE DISCUSSED WITH PARENTS, INCLUDING THE POSSIBILITY OF A MINIMALLY INVASIVE OPERATION. A MINIMALLY INVASIVE OPERATION IS AN OPERATION INVOLVING AS LITTLE INCISION INTO THE BODY AS POSSIBLE.

OPERATIVE MANAGEMENT*

*IN THE OPERATING ROOM

	Statements	Consensus/no consensus	Explanations for patients/ parents
15	A stable neonate with EA should preferably be operated during working hours during the week.	+	A stable baby with EA should be operated during the daytime on a weekday
16	Antibiotics should be routinely administered perioperatively.	+	
17	A central venous line should be placed before the operation.	+	
18	An arterial line should be placed before the operation.	+	
19	Tracheoscopy should be routinely performed before the operation to evaluate the fistula(s) and other tracheolaryngeal pathology.	+	Tracheoscopy (assessment of the airway) to exclude an upper fistula or other malformations should be carried out at the start of the surgery to help plan and to prevent avoidable complications during the operation.
20	Horizontal or vertical or U-shaped (Bianchi) approaches (skin incision) are viable approaches for conventional thoracotomy.	+	
21	Muscle-sparing approach is the recommended approach for conventional thoracotomy.	+	
22	Entry through the 4th intercostal space is the recommended approach for conventional thoracotomy.	+	
23	The extrapleural approach is the preferred approach for thoracotomy.	+	
24	In cases with suspected right descending aorta, a right-sided thoracic approach is the first option.	+	
25	The azygos vein should be preserved whenever possible.	-	
26	The tracheoesophageal fistula should preferably be closed by transfixing suture.	+	
27	The esophagal anastomosis should be preferably performed with absorbable sutures.	+	
28	The esophageal anastomosis should be preferably performed with interrupted sutures.	+	
29	A transanastomotic tube should be routinely inserted.	+	
30	A chest drain should be routinely placed.	-	
31	The thoracoscopic approach is a viable option.	+	

32	THE THORACOSCOPIC APPROACH SHOULD BE ONLY PERFORMED IF SUITABLE EXPERTISE IS AVAILABLE	+	MINIMAL INVASIVE SURGERY REQUIRES A TEAM OF EXPERTS, ESPECIALLY IN ANAESTHETICS - DON'T BE AFRAID TO ASK ABOUT THIS!
33	THE THORACOSCOPIC APPROACH OFFERS THE ADVANTAGE OF MAGNIFICATION COMPARED WITH THE CONVENTIONAL APPROACH.	+	
34	THE THORACOSCOPIC APPROACH OFFERS THE ADVANTAGE OF FASTER RECOVERY COMPARED WITH THE CONVENTIONAL APPROACH.	-	
35	THE THORACOSCOPIC APPROACH OFFERS THE ADVANTAGE OF BETTER COSMESIS COMPARED WITH THE CONVENTIONAL APPROACH.	+	
36	THE THORACOSCOPIC APPROACH OFFERS THE ADVANTAGE OF LESS MUSCULOSKELETAL SEQUELAE COMPARED WITH THE CONVENTIONAL APPROACH.	+	
37	THE MAXIMUM INSUFFLATION PRESSURE OF CO_2 DURING THORACOSCOPY SHOULD NOT EXCEED 5 MM HG.	+	
38	MAXIMUM DURATION OF THORACOSCOPIC OPERATION SHOULD BE 3 HOURS.	+	
39	THE THORACOSCOPIC APPROACH HAS THE DISADVANTAGE OF LONGER OPERATIVE TIME COMPARED WITH THE CONVENTIONAL APPROACH.	+	
40	THE THORACOSCOPIC APPROACH HAS A NEGATIVE PATHOPHYSIOLOGICAL IMPACT (ACIDOSIS, CEREBRAL OXYGENATION IMPAIRMENT) COMPARED WITH THE CONVENTIONAL APPROACH.	-	
41	THE THORACOSCOPIC APPROACH HAS THE DISADVANTAGE OF A HIGHER COMPLICATION RATE COMPARED WITH THE CONVENTIONAL APPROACH.	-	
42	THERE IS NO PLACE FOR ROUTINE FUNDOPLICATION IN PATIENTS WITH EA DURING THE INITIAL OPERATION.	+	REFLUX IS WHEN FOOD AND ACID FROM THE STOMACH FLOWS BACK INTO THE ESOPHAGUS. SURGERY TO STOP REFLUX SHOULD NOT BE DONE DURING THE INITIAL SURGERY (SURGICAL CONNECTION OF THE ESOPHAGUS - ANASTOMOSIS) - OTHER THAN IN EXCEPTIONAL CASES.

This part contains much surgical detail and does not necessarily need to be understood by parents. Trust the surgeon after explaining the procedure to you (refer to point 14 above) and after discussing that these recommendations are being followed.

POST-OPERATIVE MANAGEMENT*

*AFTER THE OPERATION

	STATEMENTS	CONSENSUS/NO CONSENSUS	EXPLANATIONS FOR PATIENTS/ PARENTS
43	POST-OPERATIVE VENTILATION AND RELAXATION SHOULD NOT BE ROUTINE AND SHOULD BE RESERVED FOR SELECTED PATIENTS, SUCH AS THOSE WITH TENSION ANASTOMOSIS.	+	VENTILATION AND A LONGER PERIOD OF IMMOBILISATION ARE NOT ALWAYS NECESSARY UNLESS THE ANASTOMOSIS IS MADE UNDER TENSION.
44	ROUTINE POSTOPERATIVE ANTIBIOTIC TREATMENT FOR LONGER THAN 24 HOURS SHOULD BE RECOMMENDED.	-	
45	A POSTOPERATIVE CONTRAST STUDY OF THE ESOPHAGUS SHOULD BE ROUTINELY PERFORMED BEFORE THE INITIATION OF ORAL FEEDING.	-	
46	FEEDING VIA THE TRANSANASTOMIC TUBE MAY BE ROUTINELY INITIATED AT 24 HOURS POSTOPERATIVELY.	+	24 HOURS AFTER THE OPERATION, FEEDING CAN START VIA A GASTRIC TUBE, A TUBE THAT BRINGS FOOD DIRECTLY INTO THE STOMACH.
47	ORAL FEEDING MAY BE ROUTINELY INITIATED AFTER 24 HOURS POSTOPERATIVELY.	+	AFTER 24 HOURS, THE CHILD MAY ALSO BE OFFERED SMALL QUANTITIES OF MILK ORALLY (VIA THEIR MOUTH) IF APPROPRIATE.
48	AN ANASTOMOTIC LEAKAGE SHOULD BE ROUTINELY MANAGED WITH A CHEST DRAIN.	+	A LEAK AT THE ANASTOMOSIS SHOULD FIRST BE TREATED BY DRAINING THE FLUID FROM THE CHEST.
49	AN ANASTOMOTIC LEAKAGE WITHIN THE FIRST 4 POSTOPERATIVE DAYS MAY BE CONSIDERED FOR SURGICAL REVISION.	-	A LEAK AT THE ANASTOMOSIS IS **NOT NECESSARILY** A REASON FOR FURTHER SURGERY IN THE FIRST 4 DAYS AFTER SURGERY (SEE ITEM 48).
50	A CONTRAST STUDY, TRACHEOSCOPY AND ESOPHAGOSCOPY ARE NECESSARY TO EXCLUDE A RE-FISTULA, OR MISSED UPPER POUCH FISTULA, IF SUSPECTED.	+	
51	A REFISTULA MAY BE INITIALLY MANAGED BY EITHER ENDOSCOPIC OR SURGICAL APPROACH.	+	
52	A CLINICAL CHECKLIST SHOULD BE MADE AVAILABLE INCLUDING ITEMS WHICH SHOULD BE PERFORMED BEFORE FIRST DISCHARGE (I.E. ABDOMINAL AND RENAL ULTRASOUND, RESUSITATION TRAINING FOR PARENTS/CAREGIVERS).	+	A CHECKLIST SHOULD BE MADE AVAILABLE INCLUDING FURTHER EXAMINATIONS BEFORE DISCHARGE TO RULE OUT POSSIBLE FURTHER MALFORMATIONS OR TO DIAGNOSE THEM EARLY. PARENTAL TRAINING IN RESPECT TO RESUSCITATION SHOULD ALWAYS BE PROVIDED BEFORE THE FIRST HOSPITAL DISCHARGE.

FOLLOW UP AND FRAMEWORK

FOLLOW UP

	FOLLOW UP	CONSENSUS/NO CONSENSUS	EXPLANATIONS FOR PATIENTS/PARENTS
1	THERE SHOULD BE A STRUCTURED SCHEDULE FOR LIFE-LONG FOLLOW UP.	+	THERE SHOULD BE A STRUCTURED SCHEDULE FOR LIFE-LONG FOLLOW UP/AFTER CARE.
2	THERE SHOULD BE AN INTERDISCPLINARY FOLLOW UP PROGRAMME INCLUDING SURGERY, GASTROENTEROLOGY, PULMONARY, NUTRITION COUNSELLING, OTOLARYNGOLOGY AND OTHERS, WITH ONE SPECIALIST LEADING.	+	FOLLOW-UP/AFTER-CARE SHOULD INVOLVE A MULTI-DISCIPLINARY TEAM LED BY ONE PARTICULAR SPECIALIST (TYPICALLY THE PAEDIATRIC SURGEON BUT COULD BE, FOR INSTANCE, A SPECIALIST PAEDIATRICIAN).
3	ANTACID MEDICATION SHOULD BE ROUTINELY ADMINISTERED TO ALL PATIENTS AFTER CORRECTION OF EA	+	
4	PROTON PUMP INHIBITORS (PPIS) SHOULD BE USED FOR ANTACID PROPHYLAXIS IN EA PATIENTS.	+	PPI (MEDICATION THAT IS SWALLOWED TO TREAT REFLUX) SHOULD BE GIVEN AS A PREVENTIVE MEASURE.
5	ANTACID MEDICATION SHOULD BE ROUTINELY ADMINISTERED FOR 12 MONTHS AFTER CORRECTION OF EA, ALTHOUGH IT IS NOTED THAT THE EVIDENCE BASE IS LIMITED.	-	ERNICA RECOMMENDS ONLY 6 MONTHS PPI TREATMENT INSTEAD OF 12 MONTHS.
6	ANTACID THERAPY SHOULD BE TAPERED AT THE END OF PROPHYLAXIS.	+	WHEN THE INITIAL PREVENTIVE TREATMENT FOR REFLUX IS STOPPING THIS SHOULD BE SLOWLY REDUCED.
7	ANASTOMOTIC STRICTURE SHOULD BE DIAGNOSED BY EITHER CONTRAST STUDY AND/OR ENDOSCOPY.	+	
8	ANASTOMOTIC STRICTURE SHOULD BE MANAGED BY BALLOON OR SEMIRIGID DILATATION.	+	STRICTURES DO NOT DILATE BY DRINKING OR EATING AND SHOULD BE STRETCHED BY ONE OF THE METHODS INDICATED BELOW. THERE ARE TWO DIFFERENT WAYS THAT ESOPHAGEAL DILATATION CAN BE DONE. IN MOST CASES, A SMALL BALLOON IS INSERTED INSIDE THE STRICTURE. THE BALLOON INFLATES TO STRETCH OPEN THE STRICTURE, AND THE BALLOON IS REMOVED. ANOTHER WAY IS TO PASS A TAPERED ELASTIC TUBE CALLED A BOUGUE INTO THE ESOPHAGUS TO THE STRICTURE. DURING THE DILATATION PROCEDURE, THE DOCTOR WILL USE SEVERAL TUBES, ONE AFTER THE OTHER, AND THE TUBES GET WIDER EACH TIME. THIS GRADUALLY STRETCHES THE STRUCTURE.
9	THE DEFINITION OF RECURRENT ANASTOMOTIC STRICTURE IS 3 ANASTOMOTIC STRICTURE RELAPSES REQUIRING DILATATION	+	
10	THE MAXIMUM NUMBER OF ESOPHAGEAL DILATATIONS FOR RECURRENT ANASTOMOTIC STRICTURES UNTIL A FUNDOPLICATION SHOULD BE CONSIDERED IS 5.	+	IF THERE ARE MORE THAN 5 DILATATIONS PERFORMED, THE BENEFIT OF ANTI-REFLUX SURGERY SHOULD BE CONSIDERED – FUNDOPLICATION. THIS SURGICAL PROCEDURE AIMS TO TIGHTEN THE VALVE AT THE TOP OF THE STOMACH, TO STOP STOMACH ACID FROM RISING UP.
11	TOPICAL APPLICATION OF MITOMYCIN C SHOULD BE RECOMMENDED AS AN OPTION IN PATIENTS WITH RECURRENT STRICTURES	-	

12	Intralesional/systemic steroids should be recommended as an option in patients with recurrent strictures.	-	
13	Customized stents /indwelling balloons should be recommended as an option in patients with recurrent strictures.	+	
14	24-hour-pH- or pH-impedance monitoring should be routinely used for monitoring children and adolescents with EA according to a specific schedule.	+	
15	24-hour-pH- or pH-impedance monitoring should be routinely performed at time of discontinuation of antacid therapy.	+	
16	At least two additional pH studies should be routinely performed until transition.	-	
17	Endoscopies of the upper gastrointestinal tract should be routinely used for monitoring children and adolescents with EA according to a specific schedule.	+	
18	Endoscopies of the upper gastrointestinal tract should be routinely performed at 1 year.	+	Esophageal endoscopies should be routinely done at the age of one year, even if there are no problems. A small flexible camera is passed via the mouth to look at the esophagus and stomach under general anaesthetic.
19	At least two additional endoscopies of the upper gastrointestinal tract should be routinely performed until transition.	+	Up until the age of 18, at least **two** such endoscopies should be carried out - even if there are no apparent problems
20	Lung function tests should be routinely used for monitoring children and adolescents with EA according to a specific schedule.	+	Lung function should be checked regularly (see also point 2 above). [And, though not included in this clinical statement, continued into adulthood as part of a life-long surveillance programme; see point 24 below].
21	Contrast study of the upper gastrointestinal tract should be routinely used for monitoring children and adolescents with EA according to a specific schedule.	-	
22	Bronchoscopy should be routinely used for monitoring children and adolescents with EA according to a specific schedule.	-	

23	THE FOLLOWING ARE POTENTIAL INDICATORS FOR FUNDOPLICATION: (I) RECURRENT ANASTOMOTIC STRICTURES (II) POORLY CONTROLLED GERD DESPITE MAXIMUM PPI THERAPY (III) LONG TERM DEPENDENCY ON TRANSPYLORIC FEEDING (IV) CYANOTIC SPELLS	+	AN ANTI-REFLUX OPERATION (FUNDOPLICATION) MAY BE NECESSARY IN THE FOLLOWING SITUATIONS: 1. PERSISTENT REUCRRING STRICTURES 2. POORLY CONTROLLED REFLUX DESPITE MAXIMUM PERMITTED DOSES OF PPI MEDICATION 3. LONG TERM TUBE-FEEDING 4. BREATHLESSNESS AND/OR INTERRUPTED BREATHING (BLUE SPELLS)
24	ADULT EA PATIENTS NEED SURVEILLANCE AS PER ESPGHAN GUIDELINES: (I) ROUTINE ENSOSCOPY (WITH BIOPSIES IN 4 QUADRANTS AT GASTROESOPHAGEAL JUNCTION AND ANASTOMOTIC SITE) AT TIME OF TRANSITION INTO ADULTHOOD AND EVERY 5 TO 10 YEARS. (II) ADDITIONAL ENDOSCOPY IF NEW OR WORSENING SYMPTOMS DEVELOP (III) IN PRESENCE OF BARRETTT AS PER CONSENSUS RECOMMENDATIONS	+	AFTERCARE AND/OR SURVEILLANCE PROGRAMMES SHOULD EXTEND INTO ADULTHOOD AND THROUGHOUT THE LIFE OF EACH PATIENT (AS PART OF THE ESPGHAN GUIDELINES): 1. ROUTINE ENDOSCOPY OF THE ESOPHAGUS WITH TISSUE SAMPLES TAKEN FOR INVESTIGATION EVERY 5-10 YEARS 2. OCCASIONAL ENDOSCOPY FOR NEW OR WORSENING SYMPTOMS (E.G. DYSPHAGIA) 3. IF BARRETT'S IS IDENTIFIED, REFERENCE SHOULD BE MADE TO THE SPECIFIC BARRETT'S GUIDELINES
25	QUALITY OF LIFE ASSESSMENT USING A VALIDATED INSTRUMENT SHOULD BE OFFERED DURING FOLLOW-UP IN CHILDREN	+	QUESTIONS REGARDING QUALITY OF LIFE SHOULD BE DISCUSSED AS PART OF THE AFTERCARE PROGRAMME.

VARIA

	VARIA	CONSENSUS/NO CONSENSUS	EXPLANATIONS FOR PATIENTS/PARENTS
26	WHEN EA IS SUSPECTED, REFERRAL TO ANTENATAL MULTIDISCPLINARY COUNSELLING IN A SPECIALISED CENTRE SHOULD BE MADE.	+	A PREGNANT WOMAN SUSPECTED OF HAVING A BABY WITH ESOPHAGEAL ATRESIA SHOULD BE REFERRED TO A SPECIALISED CENTRE THAT HAS COMPETENCE IN ESOPHAGEAL ATRESIA SURGERY AND FOLLOW-UP.
27	THERE SHOULD BE A MINIMUM AVERAGE CASELOAD OF 5 NEW EA PATIENTS PER YEAR TO MEET THE REQUIREMENT OF A SPECIALISED CENTRE.	+	AN EXPERT CENTRE SHOULD SEE A MINIMUM OF FIVE NEW BABIES WITH ESOPHAGEAL ATRESIA EACH YEAR.
28	EA PATIENTS SHOULD BE OPERATED ON AND TREATED IN SPECIALISED CENTRES WITH A MULTIDISCPLINARY TEAM WITH FOLLOW UP INCLUDING TRANSITION	+	ALL EA PATIENTS SHOULD BE OPERATED AND TREATED IN AN EXPERT CENTRE. ONLY A CLINIC THAT OFFERS A MULTI-DISCIPLINARY STRUCTURED FOLLOW-UP PROGRAMME AND SUPPORT DURING TRANSITION FROM CHILD TO ADULT CARE, IN ADDITION TO THE SURGICAL CARE SHOULD BE CALLED A SPECIALISED CENTRE.
29	PARENTS OF EA PATIENTS SHOULD BE RECOMMENDED TO BE INVOLVED IN PARENT AND PATIENT SUPPORT GROUPS AS EARLY AS POSSIBLE	+	PARENTS SHOULD BE STRONGLY RECOMMENDED TO CONTACT A PATIENT SUPPORT GROUP AS EARLY AS POSSIBLE.

Even Esophageal Atresia Type 3b / Type C (with lower fistula) is considered by international experts to be a complex malformation, and should be referred to an expert centre. All Esophageal Atresia patients should always be treated in an expert centre, despite the opinion of some surgeons that only Type 2 / Type A (no fistula and hence long-gap) should be considered as complex patients. The above international consensus statements - for the first time - clearly contradict such a position.

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