

# Esophageal Atresia Follow-Up Program Guideline

# **Objective**

To provide a comprehensive, evidence-based resource for clinicians managing long-term follow-up of esophageal atresia (EA) patients in the outpatient arena using standardized guidelines

# **Target Population**

Children and adolescents who have been treated for Esophageal Atresia (all types) and seek follow-up care at Cincinnati Children's Hospital Medical Center.

#### Evidence-Based Care Recommendations

1. It is recommended that patients with Esophageal Atresia (EA) are followed and monitored by a multidisciplinary team per the outlined schedule below (Grasso, 2025 [4a]; Krishnan, 2023 [5a]; Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Kumari, 2019 [4a]; Mirra, 2017 [1b]; Krishnan, 2016 [5a]) with one lead discipline specialist (Dingemann, 2021 [5a]; Dingemann, 2020 [5a]):

Recommendation Strength Moderate

		Esc	ophageal Follow	Up Sch	edule *							
	1 - 3 months	3 - 6 Months	Yearly	Age 1	Age 5	Age 6	Age 10	Age 15	Transition			
	Post	post Discharge		year	years	Years	Years	Years				
	Discharge	until age 1 year										
**Lead Discipline (i.e. Pediatric Surgery)												
Follow-Up Visit	1 X	зX	зX	X								
			Pulmona	ry Med								
Follow-Up Visit	2X		As indicated	Х		Х						
Flex Bronchoscopy				4X	·							
PFT's						5 <b>X</b>						
ENT												
Follow-Up Visit	2X											
MLB				4X								
			Gastroent	erology								
Follow-Up Visit	<sup>2</sup> X		As indicated	7 <b>X</b>	<sup>7</sup> X		<sup>7</sup> X	7 <b>X</b>	<sup>7</sup> X			
EGD with Biopsy				4X	еX		<sup>6</sup> Х	eХ	eХ			
Esophagram	2 <b>X</b>											
			Nutrit	ion								
Follow-Up Visit	<sup>2</sup> X	εX	εX	ВX	8X		8X	<sup>8</sup> X	8X			
Urine Na	2X											
Ferritin, CBC, Renal, Vitamin D	2 <b>X</b>											

Patients can be seen by any or all disciplines as patient's condition warrants (LC). \*\* Lead Discipline at CCHMC is Surgery\*

- Follow up with pediatric surgery 1 month post discharge (Monje Fuente, 2023 [4a]; Dingemann, 2021 [5a]).
- Establish baseline at 1 to 3 months post discharge with the following: Pulmonary, ENT, GI, and Nutrition (Krishnan, 2023 [5a]; Monje Fuente, 2023 [4a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Menzies, 2017 [3b]; Mirra, 2017 [1b]; Krishnan, 2016 [5a]; Presse, 2016 [4b]) coordinated by one Lead Discipline (Dingemann, 2021 [5a]; Dingemenn, 2020 [5a]). Perform esophagram (Pai, 2025 [3b]; Yasuda, 2020 [4a]; Krishnan, 2016 [5a]; Yasuda, 2024 [4a]), and obtain urine and blood samples for lab studies at 3 months (Local Consensus, 2025 [5]).
- 3. Follow up with Lead Discipline (Dingemann, 2021 [5a]; Dingemann, 2020 [5a]) every 3 to 6 months until age 1 year (Monje Fuente, 2023 [4a]; Dingemann, 2021 [5a]) and yearly thereafter until transition (Local Consensus, 2025 [5]; Monje Fuente, 2023 [4a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Mirra, 2017 [1b]; Krishnan, 2016 [5a]). Lead visits should include height, weight, head circumference (as appropriate) (Local Consensus, 2025 [5]; Krishnan, 2023 [5a]; Maan, 2021 [3b]; Koumbourlis, 2020 [5a]; Kumari, 2019 [4a]; Krishnan, 2016 [5a]), Dysphagia screening (Stewart, 2024 [1b]; Krishnan, 2023 [5a]; Salcedo Arroyo, 2023 [4a]; Soyer, 2017 [3b]; Coppens, 2016 [4a]; Krishnan, 2016 [5a]) and Quality of Life Assessment (Stewart, 2024 [1b]; Krishnan, 2023 [5a]; Ardenghi, 2022 [4b]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Kumari, 2019 [4a]; Dellenmark-Blom, 2018 [2a]).
- Recommend an EGD/MLB/flexible bronchoscopy be completed at 1 year of age for baseline (Grasso, 2025 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann 2020 [5a]; Koumbourlis, 2020 [5a]; Mirra, 2017 [1b]; Krishnan, 2016 [5a]; Yasuda, 2024 [4a])
- 5. Follow up with Pulmonary visits as indicated (Grasso, 2025 [4a]; Krishnan, 2023 [5a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Donoso, 2020 [4b]; Koumbourlis, 2020 [5a]; Dittrich, 2017 [3b]; Mirra, 2017 [1b]; Cartabuke, 2016 [4a]; Krishnan, 2016 [5a]; Schneider, 2014 [4a]) baseline PFT starting at age 6 years and ongoing as indicated (Local Consensus, 2025 [5]; Krishnan, 2023 [5a]; Tokarska, 2022 [5a]; Dingemann, 2020 [5a]; Donoso, 2020 [4b]; Koumbourlis, 2020 [5a]; Mirra, 2017 [1b]; Cartabuke, 2016 [4a]).
- 6. Repeat EGD with biopsy (Grasso, 2025 [4a]) every 5 years (Monje Fuente, 2023 [4a]; Tokarska, 2022 [5a]; Krishnan, 2016 [5a]; Yasuda, 2024 [4a]), and at time of transition (Krishnan, 2023 [5a]; Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Krishnan, 2016 [5a]).
- 7. Follow up with GI (Grasso, 2025 [4a]; Monje Fuente, 2023 [4a]) as indicated and; if patient continues receiving PPI (Krishnan, 2023 [5a]; Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemenn, 2020 [5a]), receives enteral feeds or prior to scopes (Local Consensus, 2025 [5]). If patient continues PPI due to GERD symptoms, schedule pH monitoring as directed by GI (Local Consensus, 2025 [5]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Krishnan, 2016 [5a]).
- 8. Follow up with Nutrition (Grasso, 2025 [4a]; Traini, 2022 [4b]; Birketvedt, 2020 [4b]; Menzies, 2017 [3b]; Krishnan, 2016 [5a]; Presse, 2016 [4b]; Schneider, 2014 [4a]; Wang, 2022 [4b]) every 3 months until age 1 year old (Krishnan, 2023 [5a]; Dingemann, 2021 [5a]), yearly to age 5 years old, then every 5 years up to transition (Local Consensus, 2025 [5]).

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#### General

It is recommended that patients with EA follow the established schedule (as outlined above) for baseline and ongoing evaluation beginning at the age first seen in the clinic (Local Consensus, 2025 [5]).

Recommendation Strength Moderate

It is recommended that an esophagram be completed around 3 months post surgical repair (Pai, 2025 [3b]; Yasuda, 2020 [4a]; Yasuda, 2024 [4a]) and then an esophagogastroduodenoscopy (EGD), microlaryngoscopy bronchoscopy (MLB), and/or flexible bronchoscopy at 1 year of age for baseline (Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Mirra, 2017 [1b]; Krishnan, 2016 [5a]).

Recommendation Strength Moderate

 It is recommended that patients with EA are monitored for height, weight, (Traini, 2022 [4b]: Maan. 2021 [3b]: Birketvedt. 2020 [4b]: Presse. 2016 [4b]: Schneider. 2014 [4a]) and head circumference up to age two years, then height and weight at least yearly thereafter (Krishnan, 2023 [5a]; Koumbourlis, 2020 [5a]; Kumari, 2019 [4a]; Krishnan, 2016 [5a]; Wang, 2022 [4b]).

Recommendation Strength Moderate

- Note: Height, weight, and head circumference (infants only) are the gold standards to measure nutrition and growth in infants, children, and adolescence (Kumari, 2019 [4a]; Krishnan, 2016 [5a]).
- It is recommended that quality of life screenings be assessed (Stewart, 2024 [1b]; Krishnan, 2023 [5a]; Salcedo Arroyo, 2023 [4a]; Ardenghi, 2022 [4b]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Kumari, 2019 [4a]; Krishnan, 2016 [5a]) beginning at age two years old (Dellenmark-Blom, 2018 [2a]) and repeated on a yearly basis (Local Consensus, 2025 [5]).

Recommendation Strength Moderate

- Note: The Esophageal Atresia-Quality-of-Life questionnaire (EA-QOL-questionnaire) can be used to assess QoL (Dellenmark-Blom, 2018 [2a]).
- It is recommended that dysphagia screening be assessed (Stewart, 2024 [1b]: Krishnan, 2023 [5a]; Salcedo Arroyo, 2023 [4a]; Soyer, 2017 [3b]; Coppens, 2016 [4a]; Krishnan, 2016 [5a]) beginning at age 6 months and repeated on a yearly basis (Local Consensus, 2025 [5]).

Recommendation Strength Moderate

- Note: The Pediatric Eating Assessment Tool-10 (pEAT-10) can be used to assess dysphagia (Stewart, 2024 [1b]; Salcedo Arroyo, 2023 [4a]; Soyer, 2017 [3b]; Cartabuke, 2016 [4a]).
- 7. It is recommended that if any EA patient is experiencing any unexpected signs or symptoms at scheduled followed up, the patient should be referred to the appropriate discipline for further assessment, management and/or treatment as the condition warrants (Local Consensus, 2025 [5]).

Recommendation Strength Moderate

#### **Pulmonary**

It is recommended that a dynamic flexible bronchoscopy procedure be done at 1 year of age for baseline (Koumbourlis, 2020 [5a]; Mirra, 2017 [1b]), then repeated as patient's condition warrants (Krishnan, 2023 [5a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Mirra, 2017 [1b]).

Recommendation Strength Moderate

- Note: The dynamic flexible bronchoscopy is the gold standard for the assessment of tracheobronchomalacia (Koumbourlis, 2020) [5a]). Rigid bronchoscopies tend to underestimate the degree of tracheal collapse because it can "stent" the airway (Koumbourlis, 2020 [5a]).
- 9. It is recommended that all patients with EA be followed by pulmonary at 1 year of age and ongoing as indicated (Krishnan, 2023 I5al; Tokarska, 2022 I5al; Dingemann, 2021 I5al; Donoso, 2020 [4b]; Koumbourlis, 2020 [5a]; Dittrich, 2017 [3b]; Mirra, 2017 [1b]; Cartabuke, 2016 [4a]; Krishnan, 2016 [5a]; Schneider, 2014 [4a]).

Recommendation Strength Moderate

- Note: Due to the presence of tracheomalacia in esophageal atresia-tracheoesophageal fistula (EA-TEF), patients are predisposed to other respiratory complications (Dingemann, 2021 [5a]; Koumbourlis, 2020 [5a]; Mirra, 2017 [1b]; Cartabuke, 2016 [4a]).
- 10. It is recommended that pulmonary function tests (PFTs) be conducted in children with EA at 6 years of age or when developmentally able and repeated as indicated (Krishnan, 2023 [5a]; Tokarska, 2022 [5a]; Dingemann, 2020 [5a]; Donoso, 2020 [4b]; Koumbourlis, 2020 [5a]; Mirra, 2017 [1b]; Cartabuke, 2016 [4a]).

Recommendation Strength Moderate

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#### Ear, Nose, and Throat (ENT)

11. It is recommended that patients with EA are seen by an ear, nose and throat (ENT) specialist (Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Krishnan, 2016 [5a]) at 3 - 6 months after discharge for baseline assessment and then follow up as needed (Local Consensus, 2025 [5]; Koumbourlis, 2020 [5a]; Krishnan, 2016 [5a]).

Recommendation Strength Moderate

#### Gastrointestinal

12. It is recommended that all patients with EA follow up with gastroenterology (GI) initially as indicated (Grasso, 2025 [4a]; Monie Fuente, 2023 [4a]), and annually if patient continues receiving proton pump inhibitors (PPIs) (Krishnan, 2023 [5a]; Monje Fuente, 2023 [4a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Krishnan, 2016 [5a]; Presse, 2016 [4b]), receives enteral feeds, or prior to scopes (Local Consensus, 2025 [5]).

Recommendation Strength Moderate

13. It is recommended that EA patients receive PPI therapy for one year post-repair (Dimitrov, 2024 [1b]; Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Righini Grunder, 2019 [4b]; Krishnan, 2016 [5a]).

Recommendation Strength Moderate

14. It is recommended, if a patient continues with reflux or GERD, pH monitoring be determined by the gastroenterologist (Grasso, 2025 [4a]; Dimitrov, 2024 [1b]; Tang, 2024 [4a]; Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Righini Grunder, 2019 [4b]; Krishnan, 2016 [5a]; Yasuda, 2024 [4a]).

Recommendation Strength Moderate

- Note: There was lack of consistent evidence and expert consensus regarding pH monitoring (Grasso, 2025 [4a]; Dimitrov, 2024 [1b]; Tang, 2024 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Righini Grunder, 2019 [4b]; Krishnan, 2016 [5a]; Yasuda, 2024 [4a]).
- 15. It is recommended that patients are evaluated with an esophagoscopy (EGD) and biopsy at completion of PPI (Grasso, 2025 [4a]) and every 5 years until transition to adult care (Tang, 2024 [4a]; Krishnan, 2023 [5a]; Monje Fuente, 2023 [4a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Krishnan, 2016 [5a]).

Recommendation Strength Moderate

# **Nutrition**

16. It is recommended that patients are evaluated by nutrition therapy (Krishnan, 2023 [5a]; Traini, 2022 [4b]; Dingemann, 2021 [5a]; Birketvedt, 2020 [4b]; Menzies, 2017 [3b]; Krishnan, 2016 [5a]; Presse, 2016 [4b]; Schneider, 2014 [4a]; Wang, 2022 [4b]):

Recommendation Strength Moderate

- every three months until 1 year of age (Dingemann, 2021 [5a])
- yearly until 5 years of age
- every 5 years until transition to adult care
- upon referral for baseline evaluation (Local Consensus, 2025 [5]).
- Note: The patient may be seen by the dietician at any time if nutritional concern(s) are raised during communication with the family (Krishnan, 2023 [5a]; Traini, 2022 [4b]; Dingemann, 2021 [5a]; Birketvedt, 2020 [4b]; Presse, 2016 [4b]).
- 17. It is recommended the nutrition assessment includes the following laboratory tests:

Recommendation Strength Moderate

- urine sodium at 3 months post-discharge for baseline (Local Consensus, 2025
- complete blood count (CBC) and vitamin D (Krishnan, 2023 [5a]; Birketvedt, 2020 [4b]); iron (Fe) studies (Krishnan, 2023 [5a]; Traini, 2022 [4b]; Birketvedt, 2020 [4b]); and renal panel at 3-months post-discharge for baseline and ongoing as indicated (Local Consensus, 2025 [5]).

#### Background

Esophageal atresia (EA) is a common congenital anomaly affecting neonates globally. With significant advancements in surgical techniques and postoperative care, EA is now recognized not only as a neonatal surgical concern but as a lifelong medical condition. Many individuals with EA continue to experience complications such as gastroesophageal reflux disease (GERD) and chronic respiratory symptoms, which can significantly impact their quality of life.

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Historically, the management of EA has been fragmented, with limited systematic follow-up during childhood and an absence of structured transition protocols into adult care. In response, professional organizations have collaborated to develop consensus-based guidelines aimed at establishing comprehensive, long-term follow-up strategies. However, these guidelines have often lacked clearly defined time frames for implementation.

This project seeks to address these gaps by developing evidence-based, multidisciplinary follow-up guidelines with specific, time-bound recommendations tailored to the unique needs of EA patients. An extensive literature review was conducted and evidence gathered from key specialties, including Surgery, Pulmonology, Otolaryngology (ENT), Gastroenterology, and Nutrition. Where peer-reviewed literature provided specific time frames, these were discussed and adopted when appropriate. In areas lacking definitive time frames, these were established through expert consensus following open discussion and review of the literature.

The implementation of standardized care with a multidisciplinary approach within regular timeframes facilitates earlier detection of long-term complications, particularly those involving the respiratory and gastrointestinal systems. These guidelines are intended to support the transition to oral feeding, improve the overall quality of care, and minimize the use of unnecessary medications and procedures.

#### **Clinical Questions**

Clinical Question for the guideline:

P (population/problem): In pediatric, adolescent, and young adult patients with esophageal atresia

I (intervention): does standardized follow-up care

C (comparison): compared to pre-standardization, lead to

O (outcome): patient being followed at regular intervals to detect and intervene on issues earlier, improving outcomes and

quality of life and easing transition to adult care?

# Target Users for the Recommendations

Pediatric Surgery, Pulmonary, Gastroenterology, Otolaryngology, and Nutrition

# **Evidence Syntheses** (including <u>Dimensions for Judging Recommendation Strength (See Appendix)</u>)

# Evidence Synthesis for Care Recommendation 1: Multidisciplinary Clinic Follow-up Schedule

Surgical repair of EA has been feasible for decades; however, evidence-based follow-up standards have not been universally adopted. Current guidelines and consensus statements from the European Society for Pediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN), the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN), the European Reference Network for Rare Inherited Congenital Anomalies (ERNICA), and the International Network of Esophageal Atresia (INoEA) emphasize the necessity of standardized, multidisciplinary follow-up care to enhance outcomes and quality of life for individuals with EA (Krishnan, 2023 [5a]; Flatres, 2022 [3a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Krishnan, 2016 [5a]).

A comprehensive, long-term follow-up algorithm with structured time frames has been proposed, incorporating evidencebased recommendations for ongoing assessments across surgical, otolaryngology (ENT), pulmonary, gastrointestinal, and nutritional domains. Early complications and co-morbidities significantly influence long-term quality of life. Common sequel includes but not limited to abnormal lung function, restrictive ventilatory defects (Dimitrov, 2024 [1b]), poor weight gain, GERD, and dysphagia (Dingemann, 2020 [5a]; Krishnan, 2016 [5a]).

These complications are not confined to early childhood but often persist in adolescence and adulthood (Dingemann, 2020) [5a]; Dittrich, 2017 [3b]; Krishnan, 2016 [5a]). Multidisciplinary care involving simultaneous input from various specialists has been shown to reduce the number of follow-up visits, enhance communication, and significantly improve patient continuity of Care (Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]).

Early and frequent follow-up during the first five years of life is critical for monitoring growth and neurodevelopment (Dingemann, 2021 [5a]; Maan, 2021 [3b]; Kumari, 2019 [4a]). Continued support through late childhood and adolescence is essential due to the profound impact on patients' and families' quality of life and mental health (Krishnan, 2023 [5a]; Koumbourlis, 2020 [5a]; Dittrich, 2017 [3b]). Adolescents and families require ongoing education and support to ensure effective surveillance and a smooth transition to adult care (Krishnan, 2023 [5a]).

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Multidisciplinary care also offers economic benefits by reducing travel costs, recurrent illnesses, co-pays, and lost workdays (Monje Fuente, 2023 [4a]). Implementing standardized, time sensitive follow-up protocols can help ensure more equitable and effective care for all patients with EA.

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Care Recommendation 1										
1. Safety versus Harm	⊠ Safe >			Balance	d Safety & H	□ Safe	☐ Safe < Harm			
2. Clinically Effective / Benefits Patient	Benefice     Benefice	ial/Effe	ctive	□ N	Neutral E	Effect or Ben	efit	□ Ineff	ecti	ve/No Benefit
3. Adherence (Burden for staff/patient/family; Access to care)	☐ Low Burden			☑ Moderate/Neutral Burden				☐ High Burden		
4. Cost (Cost for organization and/or patient/family)	☐ Cost–Effective				Cost-Ne	utral	☐ Cost	t–Pr	ohibitive	
5. Impact on quality of life, morbidity, or mortality	□ Positive	e Impact	t		/loderate	e/Neutral Im <sub>l</sub>	□ Neg	ative	e Impact	
6. Directness of Evidence	□ Directly	Relate	d		Somewh	at Related		☐ Indirectly Related		
7. Grade of the Body of Evidence	□ High ⊕⊕⊕	Ф	□ M €	oder ⊕⊕		⊠ Low ⊕⊕○0		Very Lov		□ Consensus ○○○○
Overall Strength of the Recommend	ation:	□ Str	ong	_	⊠ Mo	derate		eak		Consensus

# **Evidence Synthesis for Care Recommendation 2: Consultation**

Patients diagnosed with EA should be integrated into a structured, multidisciplinary follow-up schedule based on age at the time of hospital discharge or referral to a specialized clinic. This approach ensures continuity of care and addresses the complex; long-term needs associated with the condition. If any recommended evaluations or interventions are missed at the suggested age, the care schedule should be implemented (with catch-up assessment(s)) as soon as appropriate to ensure comprehensive management (Local Consensus, 2025 [5]). The designated lead service, identified by the multidisciplinary team, assumes responsibility for coordinating care and guiding families through the long-term management algorithm, ensuring that all aspects of the patient's health are addressed in a timely and cohesive manner (Local Consensus, 2025 [5]).

#### Care Recommendation 2

1. Safety versus Harm	⊠ Safe > Harm				Balance	d Safety & H	□ Safe	< Harm		
2. Clinically Effective / Benefits Patient	☑ Beneficial/Effective				leutral l	Effect or Ben	☐ Ineffective/No Benefit			
3. Adherence (Burden for staff/patient/family; Access to care)	☐ Low Burden			☑ Moderate/Neutral Burden				☐ High Burden		
4. Cost (Cost for organization and/or patient/family)	☐ Cost–Effective			⊠ (	Cost-Ne	eutral	☐ Cost–Prohibitive			
5. Impact on quality of life, morbidity, or mortality	☑ Positive Impact			□١	∕loderat	e/Neutral Im	□ Nega	tive Impact		
6. Directness of Evidence	□ Directly		d	☐ Somewhat Related				☐ Indirectly Related		
7. Grade of the Body of Evidence			Moderate B⊕⊕O		□ Low ⊕⊕○(		Very Low ⊕OOC			
Overall Strength of the Recommendation:		□ Str	ong		⊠ Mo	derate	□ We	ak	□ Consensus	

#### **Evidence Synthesis for Recommendation 3: Esophagram**

The esophagram plays a critical role in the postoperative evaluation of patients with EA. They can identify missed or recurrent tracheoesophageal fistulas, anastomotic strictures, congenital esophageal stenosis, and extrinsic esophageal compression caused by vascular rings (Krishnan, 2016 [5a]).

For patients considered low-risk and who are asymptomatic following surgical repair, it is recommended that an esophagram be performed around 3 months postoperatively. This timing allows for the early detection of anastomotic strictures that may develop silently to ensure timely intervention if needed (Pai, 2025 [3b]; Yasuda, 2022 [4a]).

# Care Recommendation 3

1. Safety versus Harm	⊠ Safe > Harm	□ Balanced Safe & Harm	□ Safe < Harm
Clinically Effective / Benefits Patient	⊠ Beneficial/Effective	☐ Neutral Effect or Benefit	☐ Ineffective/No Benefit
Adherence     (Burden for staff/patient/family; Access to care)	□ Low Burden	⊠ Moderate/Neutral Burden	□ High Burden
4. Cost (Cost for organization and/or patient/family)		□ Cost–Neutral	☐ Cost–Prohibitive
5. Impact on quality of life, morbidity, or mortality	☑ Positive Impact	☐ Moderate/Neutral Impact	☐ Negative Impact

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6. Directness of Evidence	☑ Directly Related			☐ Somewhat Related				☐ Indirectly Related		
7. Grade of the Body of Evidence	□ High ⊕⊕⊕⊕		□ Moderate ⊕⊕⊕○			⊠ Low ⊕⊕○○		Very Low		
Overall Strength of the Recommend	ation:	□ Str	ong		⊠ Mo	derate	$\square W$	eak	□ Consensus	

#### **Evidence Synthesis for Recommendation 4: Height and Weight**

Anthropometric measurements, including height, weight, and head circumference up to age two, and height and weight, thereafter, are essential tools for monitoring growth and evaluating the nutritional status of infants, children, and adolescents. Ongoing measurements are recommended to be performed at each office visit (Krishnan, 2023 [5a]; Traini, 2022 [4b]; Kumari, 2019 [4a]; Krishnan, 2016 [5a]; Wang, 2022 [4b]).

Research indicates that children diagnosed with EA typically exhibit lower weight, height, and body mass index (BMI) compared to their healthy pediatric counterparts (Krishnan, 2023 [5a]; Maan, 2021 [3b]; Birketvedt, 2020 [4b]; Kumari, 2019 [4a]; Krishnan, 2016 [5a]; Schneider, 2014 [4a]; Wang, 2022 [4b]). Nutritional assessment is critical not only during infancy when malnutrition can affect up to 15% of infants with EA (Birketvedt, 2020 [4b]), but also during adolescence, as inadequate nutrition during this period may result in insufficient body weight persisting into adulthood (Birketvedt, 2020 [4b]; Presse, 2016 [4b]; Schneider, 2014 [4a]).

#### Care Recommendation 4

1. Safety versus Harm	⊠ Safe > H		□ Bal	lance	d Safe & Har	m	☐ Safe < Harm			
2. Clinically Effective / Benefits Patient	☑ Beneficial/Effective			☐ Ne	utral E	Effect or Ben	☐ Ineffe	ctive/No Benefit		
3. Adherence (Burden for staff/patient/family; Access to care)	☑ Low Burden			□Мо	derat	e/Neutral Bu	☐ High Burden			
4. Cost (Cost for organization and/or patient/family)				☐ Co	st–Ne	utral	☐ Cost–Prohibitive			
5. Impact on quality of life, morbidity, or mortality				☐ Moderate/Neutral Impact				□ Negat	ive Impact	
6. Directness of Evidence	□ Directly F	Related		☐ Somewhat Related				☐ Indirectly Related		
7. Grade of the Body of Evidence			Moderate ⊕⊕⊕O		⊠ Low ⊕⊕○○		Very Low ⊕OOO	□ Consensus ○○○○		
Overall Strength of the Recommend	ation:	□ Stro	ong	D	☑ Mo	derate	□ We	eak [	□ Consensus	

### **Evidence Synthesis for Recommendation 5: Quality of Life**

Routine quality of life (QoL) screening is essential for identifying patients and families who may require additional support throughout the continuum of care. Annual assessments can help healthcare teams detect potential risks to a patient's well-being early on (Krishnan, 2023 [5a]; Ardenghi, 2022 [4b]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Kumari, 2019 [4a]). The Esophageal Atresia-Quality-of-Life questionnaire is condition specific for infants, children and adolescents with EA (Dellenmark-Blom, 2018 [2a]). The EA-QoL questionnaire with sound reliability and validity, helps identify relevant clinical information that supports patients with EA specific needs (Dellenmark-Blom, 2018 [2a]).

Patients with a history of multiple esophageal dilations, low birth weight, or families experiencing significant distress are particularly vulnerable to psychological challenges (Krishnan, 2023 [5a]). Research indicates that individuals with EA often report poorer general health, lower QoL scores, and reduced vitality compared to healthy peers (Krishnan, 2023 [5a]; Salcedo Arroyo, 2023 [4a]). These diminished scores are frequently associated with increased symptoms, especially gastrointestinal rather than the number of congenital anomalies, history of preterm birth, or low birth weight (Krishnan, 2023 [5a]; Ardenghi, 2022 [4b]).

Adolescents managing chronic conditions like EA are at an increased risk of developing mental health disorders due to persistent health complications. Therefore, regular mental health and QoL screenings are recommended (Krishnan, 2023 [5a]; Ardenghi, 2022 [4b]). It is advised that these evaluations begin at age one or at the time the patient first visits the follow-up clinic (Local Consensus, 2025 [5]).

# Care Recommendation 5

Safety versus Harm	☐ Safe > Harm		☐ Safe < Harm
2. Clinically Effective / Benefits Patient	☐ Beneficial/Effective	☑ Neutral Effect or Benefit	☐ Ineffective/No Benefit
3. Adherence (Burden for staff/patient/family; Access to care)	□ Low Burden	⊠ Moderate/Neutral Burden	☐ High Burden
4. Cost (Cost for organization and/or patient/family)	□ Cost–Effective	⊠ Cost–Neutral	☐ Cost–Prohibitive

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5. Impact on quality of life, morbidity, or mortality	☐ Positive	e Impac	t		e/Neutral Im	pact	☐ Neg	☐ Negative Impact		
6. Directness of Evidence	□ Directly	☑ Directly Related ☐ Somewhat Related						☐ Indirectly Related		
7. Grade of the Body of Evidence	□ High ⊕⊕⊕	□ Moderate □ Low □ □ ⊕⊕⊕○ □ □ Hoderate □ □ Low □ □					□ Very Lo		□ Consensus ○○○○	
Overall Strength of the Recommend	ation:	□ Str	ong	⊠ Mo	derate		Weak		Consensus	
Evidence Synthesis for Recommendate  Dysphagia is a prevalent symptom among par  Coppens, 2016 [4a]). Many individuals continue to aspiration and failure to thrive. A validated dy  Consensus, 2025 [5]) to monitor changes in swalle  The Pediatric Eating Assessment Tool-10 (PE the presence and severity of dysphagia in ped	tients with continuous experience some continuous conti	EA acr ce swal creenin tion (/K 0) is a v	oss a lowin g tool rishnar	g difficulties should be a, 2023 [5a]; S ted, sympto	s beyond the incorporate alcedo Arroyo om-specific	ie ne ed ai o, 202 inst	eonatal per nd complet 23 [4a]; Coppe rument des	iod ed y ens, sign	leading to yearly (Local 2016 [4a]).	
EAT-10 has proven effective in documenting to predicting the risk of aspiration associated with	he severit	y of init	ial sy	mptoms, ev						

# Care Recommendation 6

1. Safety versus Harm	☐ Safe > Ha	rm	Balance	d Safety & H	□ Safe <	Harm	
2. Clinically Effective / Benefits Patient	Beneficial/	Effective	□ Neutral	Effect or Ben	☐ Ineffec	tive/No Benefit	
3. Adherence (Burden for staff/patient/family; Access to care)	☐ Low Burde	en	⊠ Modera	te/Neutral Bu	□ High B	urden	
4. Cost (Cost for organization and/or patient/family)	☐ Cost–Effe	ctive	⊠ Cost–N	eutral	☐ Cost–Prohibitive		
5. Impact on quality of life, morbidity, or mortality	□ Positive In	npact	☐ Modera	te/Neutral Im	☐ Negative Impact		
6. Directness of Evidence	□ Directly Representation	elated	☐ Somewl	nat Related		☐ Indirec	tly Related
7. Grade of the Body of Evidence			loderate ⊖⊕⊕O	⊠ Low ⊕⊕○0		Very Low	□ Consensus ○○○○
Overall Strength of the Recommend	ation:	Strong	⊠ Mo	derate	□ We	ak 🗆	] Consensus

#### **Evidence Synthesis for Care Recommendation 7: Referrals**

If an EA patient presents during a follow-up visit with any unexpected signs or symptoms outside of the anticipated status or developmental milestones, consult the appropriate specialty or discipline for further assessment, management, and/or treatment, as clinically indicated to improve patient care outcomes (Local Consensus, 2025 [5]).

# Care Recommendation 7

1. Safety versus Harm	☐ Safe > Harm	×	Balance	d Safety & H	□ Safe <	Harm	
2. Clinically Effective / Benefits Patient	Beneficial/Effect     Beneficial/Ef	tive 🗆	Neutral E	Effect or Ben	☐ Ineffective/No Benefit		
3. Adherence (Burden for staff/patient/family; Access to care)	☐ Low Burden	⊠	Moderat	e/Neutral Bu	☐ High Burden		
4. Cost (Cost for organization and/or patient/family)	☐ Cost–Effective	×	Cost-Ne	eutral	☐ Cost–Prohibitive		
5. Impact on quality of life, morbidity, or mortality	□ Positive Impact     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □     □	: 0	Moderat	e/Neutral Imp	☐ Negative Impact		
6. Directness of Evidence	☐ Directly Related		Somewh	at Related		Indirect     Indirect	tly Related
7. Grade of the Body of Evidence	□ High ⊕⊕⊕⊕	□ Mode		□ Low □ □ How □		Very Low ⊕OOO	⊠ Consensus ○○○○
Overall Strength of the Recommenda	ation: 🗆 Str	ong	⊠ Mo	derate	□ We	ak D	Consensus

# Evidence Synthesis for Care Recommendations 8 – 9: Pulmonary

Residual esophageal and pulmonary abnormalities are frequently observed in patients with EA and tracheoesophageal fistula (TEF). These conditions necessitate vigilant and ongoing monitoring to mitigate long-term complications such as bronchiectasis and chronic lung damage (Donoso, 2020 [4b]; Cartabuke, 2016 [4a]).

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Infancy and early childhood represent critical periods for lung development. During this time, children with EA are particularly vulnerable to lower respiratory tract infections, especially within the first year of life (Krishnan, 2023 [5a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Cartabuke, 2016 [4a]). Therefore, early and systematic evaluation of lung function is essential to establish a clinical baseline, which is crucial for optimizing long-term respiratory outcomes (Krishnan, 2023 [5a]; Dittrich, 2017 [3b]; Mirra, 2017 [1b]).

Follow-up with a pulmonologist is recommended (Koumbourlis, 2020 [5a]), along with the use of diagnostic tools such as dynamic flexible bronchoscopy within the first year of life (Local Consensus, 2025 [5]). Additional follow up with a Pulmonologist will be determined by severity of symptoms. Patients with moderate to severe tracheomalacia symptoms may warrant regular follow-up as frequently as every 6 months (Mirra, 2017 [1b]).

# **Evidence Synthesis for Care Recommendation 10: PFT**

Given the high prevalence of impaired pulmonary function among individuals with EA and TEF, early and systematic assessment of respiratory health is essential. The use of PFTs typically start at or around 6 years of age (or when developmentally able to follow directions) to establish baseline for future lung function (*Tokarska*, 2022 [5a]; Mirra, 2017 [1b]; Cartabuke, 2016 [4a]). The frequency of repeat PFTs after initial baseline is determined by Pulmonary based on clinical assessment (*Local Consensus*, 2025). Pulmonary function and respiratory morbidity tend to not improve with age in this population, highlighting the need for early preventive strategies and clinical monitoring (*Donoso*, 2020 [4b]).

#### Care Recommendation 8 to 10

1. Safety versus Harm	⊠ Safe >	Harm		□ R:	alance	d Safety & H	arm	□ Safe	< Harm
Clinically Effective / Benefits Patient	Benefice     Benefice	rtive			Effect or Ben	☐ Ineffective/No Benefit			
Adherence     (Burden for staff/patient/family; Access to care)	□ Low Burden					e/Neutral Bu	☐ High Burden		
4. Cost (Cost for organization and/or patient/family)	☐ Cost–Effective			⊠ C	ost–Ne	eutral	☐ Cost–Prohibitive		
5. Impact on quality of life, morbidity, or mortality	☑ Positive Impact			☐ Moderate/Neutral Impact				□ Nega	tive Impact
6. Directness of Evidence	□ Directly	Related	d	☐ Somewhat Related				☐ Indire	ctly Related
7. Grade of the Body of Evidence			Moderate ⊕⊕⊕O		□ Low ⊕⊕○○		Very Low	□ Consensus ○○○○	
Overall Strength of the Recommend	ation:	□ Str	ong		⊠ Mo	derate	□ We	eak	□ Consensus

# Evidence Synthesis for Care Recommendation 11: Ear, Nose, and Throat

Current clinical guidelines and consensus statements from leading organizations, including ESPGHAN, NASPGHAN, ERNICA, and INoEA underscore the importance of standardized, multidisciplinary follow-up care. This care model explicitly includes the involvement of Otolaryngology specialists, who play a critical role in the early evaluation of patients, particularly for identifying aspiration and dysphagia (*Krishnan*, 2023 [5a]; Flatres, 2022 [3a]; Dingemann, 2020 [5a]; Koumbourlis, 2020 [5a]; Krishnan, 2016 [5a]). Patients with EA will be seen at initial evaluation and then as indicated (*Local Consensus*, 2025 [5]).

### Care Recommendation 11

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Consensus		
□ Consensus		
ga ire		

Publication Date: September 29, 2025



# Evidence Synthesis for Care Recommendations 12 - 14: GI/PPI

Esophageal atresia, with or without transesophageal fistula (TEF), is associated with significant gastrointestinal morbidity that begins in early childhood and often persists in adulthood, impacting overall quality of life. Common short- and long-term complications include gastroesophageal reflux disease (GERD), peptic esophagitis, gastric metaplasia, Barrett's esophagus, anastomotic strictures (AS), feeding disorders, dysphagia, and esophageal dysmotility (Grasso, 2025 [4a]; Krishnan, 2016 [5a]).

Current clinical guidelines and consensus statements from leading organizations including ESPGHAN, NASPGHAN, ERNICA, and INoEA emphasize the importance of standardized, multidisciplinary follow-up care. This model includes active involvement of gastroenterology, who are essential in managing both short- and long-term PPI therapy, nutritional and growth concerns, and endoscopic evaluation of the esophagus (Krishnan, 2023 [5a]; Flatres, 2022 [3a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Krishnan, 2016 [5a]). Patients with EA will be evaluated initially and annually ongoing as long as they remain on PPI therapy, receive enteral feeds and continue experiencing GERD symptoms (Local Consensus, 2025 [5]).

Initiating PPI therapy during the first year of life is standard practice (Dimitrov, 2024 [1b]; Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Righini Grunder, 2019 [4b]; Krishnan, 2016 [5a]). It was reported standardized PPI monitoring led to a 47.4% discontinuation rate of anti-reflux therapy, with a reduction in discontinuation time by 24 months (Monje Fuente, 2023 [4a]). Recent studies suggest that prolonged PPI use and cumulative exposure in infants with EA may alter gut microbiota composition, potentially increasing the risk of allergic conditions such as eosinophilic esophagitis (EOE) (Dimitrov, 2024 [1b]; Tang, 2024 [4a]). Additional concerns regarding long-term PPI use remain controversial and underexplored (Krishnan, 2016 [5a]). Discontinuation of PPI at 12 months of age has been recommended with monitoring for recurrent reflux symptoms (Dimitrov, 2024 [1b]; Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Righini Grunder, 2019 [4b]; Krishnan, 2016 [5a]).

However, if PPI therapy is not discontinued by the age of one year old, it is recommended pH monitoring should be performed upon completion of PPI treatment (Dimitrov, 2024 [1b]; Tang, 2024 [4a]; Tokarska, 2022 [5a]; Krishnan, 2016 [5a]). Consensus lacked "normal" values for pH-impedance reflux parameters in both healthy or EA children and pH-impedance failed to show any predictive correlation with actual esophagitis (Yasuda, 2024 [4a]). Recent studies show that 64.3% of infants at 18 months of age exhibited acid reflux after PPI discontinuation, supporting the need for follow-up with EGD with biopsy to evaluate the potential need for restarting PPI therapy (Fiatres, 2022 [3a]). Due to lack of consistent evidence and consensus in the national and international arena, pH monitoring can be ordered based on patient symptomatology per GI. EGD with biopsy results can be used to determine if PPI discontinuation is appropriate (Local Consensus, 2025 [5]; Yasuda, 2024 [4a]).

# Care Recommendations 12 to 14

1. Safety versus Harm	□ Safe >	Harm		⊠ E	Balance	d Safe & Har	m	□ Safe	< Harm
2. Clinically Effective / Benefits Patient	☐ Benefic	ial/Effe	ctive	⊠ N	leutral l	Effect or Ben	efit	☐ Ineffe	ctive/No Benefit
3. Adherence (Burden for staff/patient/family; Access to care)	☐ Low Bu	ırden		⊠ N	Иoderat	e/Neutral Bu	rden	□ High	Burden
4. Cost (Cost for organization and/or patient/family)	☐ Cost–E	ffective		$\boxtimes$ (	Cost-Ne	eutral		☐ Cost-	-Prohibitive
5. Impact on quality of life, morbidity, or mortality	☐ Positive	e Impac	t	⊠ N	√oderat	e/Neutral Im	oact	□ Nega	tive Impact
6. Directness of Evidence	☐ Directly Related		d	Somewhat Related			☐ Indirectly Related		
7. Grade of the Body of Evidence	□ High ⊕⊕⊕	$\oplus$	⊠ Mo ⊕	oder ⊕⊕		□ Low ⊕⊕○(		Very Low ⊕OOO	
Overall Strength of the Recommend	ation:	□ Str	ong		⊠ Mo	derate	□ We	eak	□ Consensus

# **Evidence Synthesis for Care Recommendation 15: EGD with Biopsy**

Esophagoscopy with biopsy remains the gold standard for screening both peptic and eosinophilic esophagitis in patients with EA (Dingemann, 2021 [5a]; Krishnan, 2016 [5a]). Routine surveillance endoscopies with biopsy are recommended for all EA patients, including those who are asymptomatic (Grasso, 2025 [4a]; Monje Fuente, 2023 [4a]; Krishnan, 2016 [5a]; Yasuda, 2024 [4a]). Notably, esophageal mucosal abnormalities were present in up to 35% of EA patients during endoscopy, even in the absence of clinical symptoms (Krishnan, 2016 [5a]).

**6000** 

□ Weak

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□ Consensus

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**⋈** Moderate





7. Grade of the Body of Evidence

Overall Strength of the Recommendation:

While the exact timing of endoscopic evaluations varies, completing at least three endoscopic evaluations prior to transition to adult care was the most consistently recommended approach in the literature.

(Tang, 2024 [4a]; Krishnan, 2023 [5a]; Monje Fuente, 2023 [4a]; Flatres, 2022 [3a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Righini Grunder, 2019 [4b]; Krishnan, 2016 [5a]). Following PPI discontinuation, EGD with biopsy is recommended to confirm the absence of gastric reflux sequela (Krishnan, 2023 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Krishnan, 2016 [5a]). These should be performed around age 5, before age 10, and again at the time of transition to adult care (Krishnan, 2023 [5a]; Monje Fuente, 2023 [4a]; Tokarska, 2022 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Krishnan, 2016 [5a]).

Care Recommendation 15				
1. Safety versus Harm	☐ Safe > Harm	⊠ Balance	ed Safe & Harm	□ Safe < Harm
Clinically Effective / Benefits Patient	⊠ Beneficial/Effect	tive □ Neutral	Effect or Benefit	☐ Ineffective/No Benefit
3. Adherence (Burden for staff/patient/family; Access to care)	☐ Low Burden	⊠ Modera	te/Neutral Burder	n □ High Burden
4. Cost (Cost for organization and/or patient/family)	□ Cost–Effective		leutral	☐ Cost–Prohibitive
5. Impact on quality of life, morbidity, or mortality	☐ Positive Impact		te/Neutral Impact	☐ Negative Impact
6. Directness of Evidence	☐ Directly Related	I ⊠ Somew	hat Related	☐ Indirectly Related
7.0 1.60 5.1.65	☐ Hiah	☐ Moderate	⊠ Low	☐ Verv Low ☐ Consensus

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# **Evidence Synthesis for Care Recommendation 16: Nutrition assessment**

Feeding difficulties, compounded by the burden of comorbidities and repeated surgical interventions, can significantly impair nutritional status in children born with EA, often leading to failure to thrive and malnutrition (Birketvedt, 2020 [4b]; Wang, 2022 [4b]). Studies report that growth in children with EA typically falls below reference norms during the first year of life, even when early nutritional support is provided (Traini, 2022 [4b]; Krishnan, 2016 [5a]; Schneider, 2014 [4a]). According to the French National Esophageal Atresia Registry, 15% of infants with EA aged 6 to 12 months were undernourished (Schneider, 2014 (4a)). Poor nutrition has been linked to delayed cognitive development, underscoring the importance of early nutritional assessment and intervention (Krishnan, 2016 [5a]; Schneider, 2014 [4a]; Wang, 2022 [4b]).

Growth retardation is notably higher in EA patients with recurrent TEF, with rates reaching 33.7% (Wang, 2022 [4b]). The pubertal growth phase presents an additional opportunity for nutritional intervention to address feeding difficulties and optimize growth outcomes (Birketvedt, 2020 [4b]; Presse, 2016 [4b]). Children with persistent feeding challenges and inadequate energy intake beyond the age of 12 are strongly associated with being underweight in adulthood (Presse, 2016 [4b]). Clinical guidelines consistently advocate for ongoing nutritional assessments throughout childhood, adolescence, and into adulthood to support optimal growth and development (Krishnan, 2023 [5a]; Dingemann, 2021 [5a]; Dingemann, 2020 [5a]; Krishnan, 2016 [5a]; Presse, 2016 [4b]; Schneider, 2014 [4a]).

#### Care Recommendation 16

1. Safety versus Harm	⊠ Safe >	Harm		☐ Balar	ncec	l Safe & Har	m	☐ Safe	e < Hai	rm
2. Clinically Effective / Benefits Patient	Benefic     Benefic	ial/Effec	tive	☐ Neut	ral E	ffect or Ben	efit	☐ Inef	fective	/No Benefit
3. Adherence (Burden for staff/patient/family; Access to care)	⊠ Low Bu	rden		□ Mode	erate	e/Neutral Bu	rden	□ Higl	n Burde	en
4. Cost (Cost for organization and/or patient/family)	☐ Cost–E	ffective		⊠ Cost-	-Ne	utral		☐ Cos	t-Proh	ibitive
5. Impact on quality of life, morbidity, or mortality	□ Positive	lmpact		☐ Mode	erate	e/Neutral Imp	oact	☐ Neg	jative li	mpact
6. Directness of Evidence	□ Directly	Related	t	□ Some	ewh	at Related		□ Indi	rectly F	Related
7. Grade of the Body of Evidence	□ High ⊕⊕⊕	$\oplus$		oderate ⊕⊕O		⊠ Low ⊕⊕○0		Very Lo		Consensus
Overall Strength of the Recommend	ation:	□ Str	ong	☒	Мо	derate	□ We	eak	□с	onsensus

# **Evidence Synthesis for Care Recommendation 17: Lab Studies**

Nutritional monitoring in patients with EA is essential due to the high prevalence of growth deficiencies in this population. A comprehensive nutritional assessment should include oral intake screening, anthropometric measurements, and biochemical markers (Birketvedt, 2020 [4b]).





Studies have shown that children with EA often do not meet the recommended dietary intake (RDI) for iron and calcium (*Traini, 2022 [4b]*). A prospective cross-sectional study of adolescents with EA found that 44% were diagnosed with vitamin D deficiency and 10% with iron deficiency, underscoring the importance of periodic laboratory evaluations (*Birketvedt, 2020 [4b]*).

To address concerns regarding poor growth in EA patients, baseline laboratory tests, including urine and blood sodium, iron, vitamin D, renal function, and complete blood count (CBC), are obtained three months post-discharge for baseline (Local Consensus, 2025 [5]). Based on local consensus among nutrition and gastroenterology teams, additional testing can be conducted as clinically indicated (Local Consensus, 2025 [5]).

# Care Recommendation 17

Safety versus Harm	⊠ Safe >	Harm		□В	Balance	ed Safe & Har	m	☐ Safe	e < Harm
2. Clinically Effective / Benefits Patient	Benefice     Benefice	cial/Effect	tive		leutral	Effect or Ben	efit	☐ Inef	fective/No Benefit
3. Adherence (Burden for staff/patient/family; Access to care)	⊠ Low Bu	ırden			/lodera	te/Neutral Bu	den	□ High	n Burden
4. Cost (Cost for organization and/or patient/family)	☐ Cost–E	ffective		⊠ C	Cost-N	eutral	_	☐ Cos	t-Prohibitive
5. Impact on quality of life, morbidity, or mortality	□ Positive	e Impact		$\square$ N	/lodera	te/Neutral Imp	oact	□ Neg	ative Impact
6. Directness of Evidence	□ Directly	/ Related		□S	Somew	hat Related	_	□ Indi	rectly Related
7. Grade of the Body of Evidence	□ High ⊕⊕⊕	Ф	□ Mc	odera		□ Low ⊕⊕○0		Very Lo ⊕OO(	
Overall Strength of the Recommenda	ation:	□ Stro	ong		⊠ Me	oderate	□ We	eak	☐ Consensus

#### **Abbreviations**

AS - Anastomotic strictures

BMI - Body mass index

CBC - Complete blood count

EA – Esophageal atresia

EA-QoL Questionnaire - Esophageal Atresia Quality of Life Questionnaire

EGD - Esophagogastroduodenoscopy

EOE - Eosinophilic esophagitis

ENT - Otolaryngology; ear, nose, and throat

ERNICA - European Reference Network for Rare Inherited Congenital Anomalies

ESPGHAN - European Society for Pediatric Gastroenterology, Hepatology and Nutrition

GA - Gestational age

GERD - Gastroesophageal reflux disease

GI – Gastrointestinal

INoEA – International Network of Esophageal Atresia

MLB - Microlaryngoscopy bronchoscopy

NASPGHAN - North American Society for Pediatric Gastroenterology, Hepatology and Nutrition

NP – Nurse practitioner

Ped – Pediatric surgery

PFT - Pulmonary function test

PPI – Proton pump inhibitor

QOL - Quality of Life

RDI - Recommended Dietary Intake

TEF - Tracheoesophageal fistula

URI - Upper respiratory infection

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# Implementation Plan

The draft follow-up table was presented to staff, including providers, for appropriateness and ease of use. Numerous iterations of the table were tested and evaluated followed by revisions based on user feedback, until the current table was published in this guideline. The table was reviewed by several current parents who provided feedback and suggestions of adding color coding for families for ease of reading and definitions for better understanding (see Appendix: Esophageal Atresia Patient Follow-up Schedule (Parental Version). These suggestions were addressed and appreciated by both staff and families.

The follow-up recommendations for Basic Care for EA patients were communicated to each individual service line separately, so feedback was obtained from support staff, NPs and Physicians. The Follow-up recommendations were approved by individual departments following discussion, feedback and revision. The completed guidelines, including recommendations, were presented at the Executive Aerodigestive Meeting for discussion and approval. Once executive approval was obtained, it was communicated to the whole Aerodigestive team at a weekly meeting. Consensus was obtained and the guidelines distributed for use in the clinic.

Esophageal clinic will be incorporated into the Aerodigestive follow up clinic. This will include the Esophageal NP. Pulmonary MD/NP, Nutrition, and GI. Office visit templates will be created for initial visits and yearly follow-up visits that include previous history, past aerodigestive scopes/testing, and future follow-up with testing. Pre-visit planning will gather information on the most recent office visits, imaging, lab testing, and patient/family concerns. An Esophageal Registry will assist with identifying who is qualified for follow up in the Esophageal clinic. Esophageal Atresia Quality of Life assessment and the Dysphagia screenings of Pediatric EAT-10 with Functional Oral Intake Scale will be completed via tablet when the families arrive in the clinic. The data will be collected and managed using REDCap, an electronic data capture tool hosted at Cincinnati Children's Hospital.

The EA Follow- up Program was developed to ensure consistent implementation of care elements as outlined in the clinical guidelines. A designated lead subspecialist is responsible for overseeing this implementation and for yearly ongoing evaluation of EA patients. At Cincinnati Children's Medical Center, this role is fulfilled by the Advance Practiced Nurse Practitioner within the Pediatric Surgery EA program. However, at other institutions, the lead may be an advanced practice provider or physician from any relevant subspecialties. The selection of the lead specialist should be determined collaboratively by the multidisciplinary team.

#### Relevant Cincinnati Children's Tools

- Family education will be developed to explain Esophageal Atresia with or without TEF and the need for long-term follow-up. This information will be updated to reflect the new follow-up schedule. Education for adolescents addressing the need to transition to adult care for lifetime screening will be developed.
- A communication template will be created to inform the primary care provider after the initial visit regarding possible feeding issues, pulmonary management of URI(s) and the need for PPI for the first year.
- An order set for EA patients discharged from the NICU will be created to reflect the initial referrals to get the follow-up process started.

#### Outcome Measures

The standardized follow-up schedule ensures all families will receive the same basic care and will provide simultaneous access to multiple specialties. This is monitored by the frequency of office visits, which specialties are seen and what testing is conducted. Outcome variables including readmission during the first year, growth and nutritional status (closely monitoring height and weight), timing for PPI discontinuation, and reduction of lung disease will be tracked. Patients will be followed at regular intervals so issues can be identified and addressed proactively. Telehealth may be an option for families who live out of town to ensure they receive the same follow-up care.

#### Potential Measures:

- Number of patients seen by multiple providers/disciplines of the multidisciplinary clinic vs individual clinic visits
- Complications identified early with screening tests and/or procedures
- Development of and detection of recurrent fistula(s)
- Improved lung and esophageal function
- Growth and development



- Dysphagia monitoring to identify if symptoms worsen or improve
- Socioeconomic factors and resource utilization (Wang, 2022 [4b])
- Detection of post-op complications within first year of surgical repair
- Early and late postoperative morbidities these frequently occur after initial repair of EA/TEF despite excellent surgical and neonatal management, and can be associated with impaired outcomes
- Ability to take oral feedings
- Access for coordinated triple scopes versus individual scopes by following standardized schedule
- Improve patient/family satisfaction by supporting the family through management of chronic illness in the **Esophageal Clinic**
- Earlier initiation of oral feeds through frequent follow-up visits during the first two years of life, providing patient and family education and support

#### **Process Measures**

- Percent of EA follow-up patients who complete one of the screening tools:
  - Dysphagia Pediatric EAT-10
  - Quality of Life, EA
  - Esophageal Risk Assessment Using the Esophageal Risk Assessment tool during pre-clinic planning, we will be able to follow if all providers are incorporating the standard follow-up guidelines and recommended testing. This pre-clinic planning will be part of the Esophageal RN workflow.
- Number of patients who are weighed, measured for height and head circumference (as appropriate).
- Number of patients who undergo EGD and biopsy as recommended by GI guidelines
- Adherence to follow up schedule (reported as a percentage)

# Search Strategies & Results

# **Search Strategy**

To select evidence for critical appraisal for this Evidence Summary, the databases below were searched using search terms, limits, filters, and date parameters to generate an unrefined, "combined evidence" database. This search strategy focused on answering the clinical questions addressed in this document and employing a combination of Boolean searching on human-indexed thesaurus terms (e.g., MeSH) as well as "natural language" searching on words in the title, abstract, and indexing terms.

Databases Searched	Search Terms	Limits, Filters, Search Dates & Parameters
MedLine     CINAHL     Cochrane     Database for     Systematic     Reviews     Google Scholar     Scopus     Hand Searching     Reference List	<ul> <li>Esophageal atresia, Esophageal atresia.mp.</li> <li>Therapeutics, treatment</li> <li>Follow up, continuity of patient care, continuity of care, long-term follow up</li> <li>After care, transition to adult care, transition of care, transition</li> <li>gastrointestinal</li> <li>Swallow study, swallowing assessment, swallow assessment</li> <li>Dysphagia, deglutition disorder</li> <li>Pulmonary, respiratory, pulmonary function test</li> <li>Quality of life, QoL</li> <li>Nutrition, nutrition assessment</li> <li>Ear nose and throat, ENT</li> <li>Vocal Cord Paralysis</li> <li>Laryngo-tracheoesophageal cleft</li> <li>PPI, proton pump inhibitor</li> <li>Follow up labs, blood test.mp. or Hematologic Tests/outpatient, ambulatory</li> </ul>	Date of Most Recent Search  • 06/27/2025  Publication Dates Searched  • 01/01/2013 to 06/27/2025  Age Groups in Evidence  • Pediatric, Adolescent, young Adult Evidence  English Language  Other Criteria  • Human  • (comparative study or controlled clinical trial or guideline or meta-analysis or multicenter study or observational study or practice guideline or randomized controlled trial or "systematic review")  Exclusions – opposite inclusion



#### Search Results & Methods

Multiple searches (electronic search engines, manual searches of citations/references) for evidence identified 789 articles. Six hundred and ninety-four articles were discarded, as they were duplicates or not related to the clinical question of interest based on title and abstract review. Ninety-four articles were reviewed in full text and critically appraised. Following full text review fifty-nine articles were excluded/discarded, as they were not related to the clinical care of EA patients or methodologically weak. Thirty-five studies/articles met the inclusion criteria above and were used in the guideline recommendations. Details and appraisal of the included studies can be obtained upon request for the Esophageal Atresia Evidence Table at EBDMInfo@cchmc.org.

# Team Members | Conflicts of Interest | External Funding

# Multidisciplinary Group / Team Members

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Special thanks to the parents who contributed with input to the development of the Esophageal Atresia Patient Follow-up Schedule (Parental Version) (see Appendix) support document.

# Conflicts of Interest:

No financial or intellectual conflicts of interest were found.



# **External Funding**

No external funding was received for the development of this guideline. Recommendations were developed through hospital funding via salaries.

# **Future Research Questions**

- 1. In children who have had esophageal atresia repair who are monitored in a multidisciplinary outpatient EA follow-up clinic does performing a dysphagia screening and/or assessment improve identification of children who may need additional screening and support?
- In children who have had esophageal atresia repair who are monitored in a multidisciplinary outpatient EA follow-up clinic does seeing a dietician on a regular basis decrease family stress?
- In children who have had esophageal atresia repair who are monitored in a multidisciplinary outpatient EA follow-up clinic does seeing a dietician on a regular basis improve patient/family quality of life?
- In children who have had esophageal atresia repair who are monitored in a multidisciplinary outpatient EA follow-up clinic, do regularly performing pulmonary function tests help decrease progression of lung dysfunction?

# Evidence-Based Clinical Care Recommendation Development Process

Recommendation statements were developed in accordance with Cincinnati Children's Evidence-Based Care Guideline Development Process (for more details, contact <u>EBDMinfo@cchmc.org</u>). The recommendations contained in this guideline were formulated by a multidisciplinary working group based on the best-available peer-reviewed evidence, patient and family values, clinical expertise, and stakeholder consensus. The team performed a systematic search and critical appraisal of the literature using the LEGEND Evidence Evaluation System (see next section below). During formulation of these recommendations, the team members have remained cognizant of controversies and disagreements over the management of these patients. Controversial issues were resolved by stakeholder and team member discussions resulting in consensus where possible and, when not possible, were offered optional approaches to care in the form of information that includes best supporting evidence of efficacy for alternative choices.

# LEGEND Evidence Evaluation System (Let Evidence Guide Every New Decision)

#### Evidence Levels of Individual Studies by Domain, Study Design, & Quality (Link to Full Table)

Individual studies are appraised for reliability, validity, and applicability, using standardized appraisal forms, to determine the Quality Level or Evidence Level (a vs b)†.

Quality Level	Definition
1 <b>a</b> <sup>†</sup> or 1 <b>b</b> <sup>†</sup>	Systematic review, meta-analysis, or meta-synthesis of multiple studies
2a or 2b	Best study design for domain
3a or 3b	Fair study design for domain
4a or 4b	Weak study design for domain
5a or 5b	General review, expert opinion, case report, consensus report, or guideline
5	Local Consensus

 $<sup>\</sup>dagger a = good guality study OR b = lesser guality study$ 

# Grade for the Body of Evidence (Link to Full Table)

The Body of Evidence (BOE) is evaluated for quantity, quality, and consistency to determine the grade of the BOE and what the impact of the BOE is on our confidence in the precision of the answer to the clinical question (and its associated recommendation statement).

Grade	Definition
High	Good quality, High-level studies with consistent results
Moderate	Good quality, Lower-level OR Lesser quality, Higher-level studies with consistent* results
Low	Good or lesser quality, Lower-level with results that may be inconsistent
Very Low	Few Good or Lesser quality, Lower-level studies that may have inconsistent results
Consensus	Local Consensus, No published evidence



#### Dimensions for Judging the Strength of the Recommendation (Link to Full Table)

- 1. Safety versus Harm
- 2. Clinically Effective / Benefits Patient
- 3. Adherence
- 4. Cost

- 5. Impact of Quality of Life, Morbidity, or Mortality
- 6. Directness of the Evidence
- 7. Grade of the Body of Evidence

#### Language and Definitions for Recommendation Strength (Link to Full Table):

Language for Strength	Definition
It is strongly recommended that It is strongly recommended that not	When the dimensions for judging the strength of the recommendation are applied (including safety/harm, health benefit, body of evidence, etc.), there is high support that benefits clearly outweigh risks and burdens. (or visa-versa for negative recommendations)
It is recommended that It is recommended that not	When the dimensions for judging the strength of the evidence are applied, there is moderate support that benefits are closely balanced with risks and burdens.
It is suggested that	When the dimensions for judging the strength of the evidence are applied,
It is suggested that not	there is weak support that benefits are closely balanced with risks and burdens.
There is insufficient evidence to make a r	ecommendation

# **Review Process**

All feedback received from internal and external reviewers was appropriately discussed and addressed by the development team.

#### Internal Review

This guideline has been reviewed against quality criteria by independent peer reviewers from Cincinnati Children's Hospital including, but not limited to, evidence methodologists, relevant subject matter experts, and/or other stakeholders who were not involved in the development process.

#### **External Review**

The guideline was externally appraised by independent peer reviewers not involved in the development process using the AGREE II instrument (Appraisal of Guidelines for Research and Evaluation II).

# Revision Process

This guideline will be removed from the Cincinnati Children's website if content has not been revised within five years of the most recent publication date. A revision of this guideline may be initiated at any point within the five-year period if evidence indicates a critical change is needed. Team members or current staff will reconvene to explore the continued validity, relevance and need of the auidelines.

# **Review History**

Date	Event	Outcome
September 29, 2025,	Original Publication	New guideline development and publication

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# For more information

About this guideline, its companion documents, or the Cincinnati Children's Evidence-Based Care Recommendation Development process, contact the Cincinnati Children's Evidence-Based Decision Making Team at <a href="mailto:EBDMInfo@cchmc.org">EBDMInfo@cchmc.org</a>.

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# **Appendix**

# **Esophageal Atresia Patient Follow-up Schedule (Parental Version)**

		Esophageal	Patient Follo	w-Up Schedu	ıle *				
	1 - 3 months	3- 6 Months	Yearly	Age 1	Age 5	Age 6	Age 10	Age 15	Transition
	Post	post Discharge	Tourty	_	Years	Years	Years	Years	Transition
		until age 1 year		year					
	Discharge								
		Lead Discipl							
***Follow-Up Visit	х	X	Х	Х	Х	х	Х	Х	Х
			Pulmonary !	Med					
Follow-Up Visit	Х		88	Х		Х	Х	Х	Х
			indicated						
****Flex Bronchoscopy				Х					
(using a scope to look at									
windpipe & lungs) PFT's						Х	Х	V	Х
(blowing test to see how well						Χ	Х	Х	X
your lungs work)									
jour tange tronny			ENT		<b>.</b>				
Follow-Up Visit	х								
****MLB				Х					
Scope to look at upper and lower									
airway)									
			Gastroentero	logy					
Follow-Up Visit	Х		as	Х	Х		Х	Х	Х
			indicated						
****EGD with Biopsy				X	Х		Х	X	Х
(scope to look through food									
pipe, stomach and upper									
intestines & get tissue sample)									
Esophagram	х								
(x-ray test with dye)			Nutrition						
E-H H- V6-14	v	V			v	I	V	V	v
Follow-Up Visit	х	Х	Х	Х	Х		Х	Х	Х
			(until age						
			5, then						
			every 5						
Urine Na	X		years)						
Ferritin, CBC, Renal, Vitamin	х								
D									
(blood work)									

<sup>\*</sup>Patients can be seen by any or all disciplines as patient's condition warrants (LC). \*\* Lead Discipline at CCHMC is Surgery

<sup>\*\*\*</sup>Follow-up visits include Dysphagia screening. Quality of Life screening, height and weight assessments
\*\*\*\*These "triple scopes" includes MLB, <u>Felx</u> Bronchoscopy and EGD procedures that are scheduled for the same time.