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A National Experience in Paediatric Achalasia: Symptom Recurrence, Reintervention, and Transition Challenges

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Abstract

Aims

To assess the long-term outcomes and quality of life (QOL) of patients who underwent intervention for esophageal achalasia in a national Paediatric centre over an 18-year period.

Methods

All patients who underwent surgical or radiological intervention after a diagnosis of achalasia between 2006 and 2024 were identified. A retrospective review of patient demographics, operative technique and Eckardt score before intervention was undertaken. Patients were contacted via telephone to re-assess QOL using Eckardt and Achalasia Scoring Questionnaire (ASQ) scoring systems.

Results

In total, 31 patients were identified and 28 were included in the analysis (15 female and 13 male). The majority 21(75%) underwent laparoscopic Heller Myotomy. The mean age at intervention was 11.6 years. The mean duration of follow-up was 7.6 years. The mean pre-operative Eckardt score was 6.9. The mean post-operative Eckardt score was 2.9. Twenty-one patients (75%) were initially symptom free for an average of 17.4 months and 7 (25%) remain symptom free to this day. Eleven (39%) required further interventions. In terms of quality of life, the current mean ASQ score is 19.4 (scoring system 10-31, with 10 representing symptom free). Of the patients who are now adults, 12 (57%) had a formal transition of care to adult services.

Conclusion

Although surgery may initially improve achalasia symptoms, it is not curative. Patients require lifelong follow-up and support to manage this challenging chronic condition. Ensuring patients are transitioned from Paediatric to adult services could help improve future QOL and tools such as ASQ could help monitor this in an outpatient setting.

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Introduction

Esophageal Achalasia (EA) is a rare neurodegenerative dysmotility disorder of the esophagus. The pathophysiology of the condition consists of incomplete or absent lower esophageal sphincter (LES) relaxation and abnormal esophageal peristalsis during swallowing[1]. The precise etiology is still unknown; however, it involves selective degeneration of inhibitory neurons of the myenteric (Auerbach's) plexus at the LES and body of the esophagus[2]. In the Paediatric age group, the incidence of EA ranges from 0.02 to 0.31 cases per 100,000, which is approximately ten times less than in the adult population[3]. Dysphagia, regurgitation or vomiting, chest pain and weight loss are commonly reported symptoms; however, atypical presentation can be seen in toddlers and infants. Delayed diagnosis of childhood EA may occur due to overlap of symptoms with other common Paediatric disorders[4]. Trisomy 21, familial dysautonomia, glucocorticoid insufficiency, congenital central hypoventilation, as well as Allgrove syndrome (triple A syndrome) are often associated with EA in children. Esophageal radiology, esophagoscopy, and high-resolution esophageal manometry are utilized to establish the diagnosis[5]. Treatment of EA aims to alleviate symptoms by reducing LES tone. This can be achieved by utilising pharmaceutical, endoscopic, radiological or surgical means. Presently laparoscopic Heller myotomy with or without a fundoplication has become the treatment of choice due to high efficacy and safety in Paediatric patients. The recurrence of symptoms after Heller myotomy has been documented with variable percentage in the literature[6,7]. In fact, there are limited data in literature regarding the impact of surgical intervention for EA on long term Quality of Life in children and gap in the literature concerning the transition of care of EA patients to adult service for long term follow-up. In our study, we aimed to evaluate long-term symptom control, need for re-intervention, and quality of life using validated scoring tools in children treated for EA over an 18-year period.

Methods

This work was approved by the ethical committee in CHI (Ref. REC440-24). The study included all patients less than16 years of age with confirmed diagnosis of EA based on clinical data, radiology and endoscopy, whether isolated or associated with problems and underwent surgical or radiological intervention for EA in the national Irish Paediatric tertiary referral centre over18 years, from 2006-2024. Exclusion criteria were uncontactable patients or those who have left the country. A retrospective review of patient's medical records was undertaken and data relating to patient demographics, medical management and investigations well as surgical technique and postoperative management with follow-up were collected. Achalasia symptoms were assessed for each patient before intervention. Eckardt score was utilized to assess the severity of EA symptoms, which were dysphagia, frequency of

retrosternal pain, weight loss and frequency of regurgitation. It assigns point to each of the 4 main symptoms; each symptom is scored from 0 (no symptoms) to 3 (severe) leading to maximum score of 12 (Supplementary Data 1). Parents of the patients were contacted by W.F and M.A who are co-authors in this paper, and consent was obtained before re-assessment of current QOL using Eckardt and Achalasia Scoring Questionnaire (ASQ) scoring systems. ASQ score is a ten-question questionnaire designed to assess the degree of difficulty in consuming multiple food items and quantifies the impact of disease on daily living (Supplementary Data 2)[8]. The phone interview was standardized to address all points in Echardt and ASQ which were interviewer- scored.

We recorded the collected data and performed analysis using Microsoft Excel 2021 version and Jamovi program version 2.3.21. Continuous variables were reported as mean ± standard deviation (SD) and range when appropriate. Categorical data were presented as the number of patients and their relative percentages. Wilcoxon test was used to compare of the mean between two non-parametric paired group. A significant p value was <0.05.

Results

Demographics, clinical presentation, and diagnostic tests

Thirty-one cases underwent operative intervention for EA after confirmation of diagnosis between 2006 to 2024 with an incidence of 1.7 cases/year. Three cases were excluded due to no response to telephone interview. Twenty-eight cases are included in the analysis with mean age at diagnosis of 11.66(1.4-15 years)/median 12, and female gender had a subtle predominance 15(54%) vs 13(46%) male. It was observed from this study that 17 patients had isolated EA wherein 11patients were associated with comorbidities (1 triple A syndrome, 3 Down's syndrome,1 Raynaud's syndrome, 1 esophageal atresia/Tracheoesophageal fistula, 1 Diabetes mellitus, 1 scoliosis and 3 with respiratory symptoms. All patients reported regurgitation, 15 had variable degrees of dysphagia, 5 developed chest pain and 3 had failure to thrive while 7 experienced weight loss. All patients except one had an upper gastrointestinal contrast study, which showed the classical bird beak or rat-tail sign with variable degrees of proximal dilated esophagus. Twenty-six (93%) had upper gastrointestinal endoscopy as part of diagnostic work up. High-resolution manometry was performed in 16(57%) of patients (Table 1).

Medical symptomatic management was attempted before surgical intervention in 15(53%) of patients which was mainly by providing patients with PPI medication and nutritional support, prior to definitive intervention. No patient had botulism toxin, nitrates or calcium channel

blockers. For each patient, the preoperative EA severity was determined via Eckardt score: the mean was 6.92 with a range from 3 to 11 and a SD 2.2 (Figure 1).

Management

The primary surgical intervention was a Laparoscopic Heller myotomy with anterior fundoplication in the majority of cases 21 (75%), open Heller myotomy with Nissen fundoplication in 4 cases (14.2%), open Heller with no antireflux procedure in 1 case (3.6%) and primary Balloon dilation in 2 cases (7.1%). No immediate postoperative complication was reported, except one case had an intraoperative mucosal breech, which was recognised and repaired intraoperatively. Most patients had a contrast swallow on the first postoperative day to ensure no oesophageal leak.

Evolution and long-term assessment of quality of life

All cases were followed in outpatient clinics by Paediatric surgery team regularly and reviewed by Gastroenterology team as needed, with mean follow-up of 7.6 years (7-218 months). Cases that were not responding to treatment plan or had persistent symptoms/recurrence were discussed in multidisciplinary team with presence of Paediatric interventional radiologist. Twenty-one cases (75%) improved initially after primary intervention with an average of 17.4 months symptom-free and 7 cases (25%) had complete resolution of their symptoms. However, 11 out 21 cases (52%), who had initial improvement, required repeated intervention due to recurrent/persistent symptoms. Interestingly, the majority of those patients had isolated EA 9 vs 3 EA with comorbidities. Nine underwent Balloon dilation, 1 a redo Heller and 1 underwent stenting of the oesophagus with a biodegradable stent.

Reassessment of Eckardt score was established via telephone contact for all patients which showed a mean of 2.9 with range from 0-5 and SD 1.4. (Figure 1). In addition, we conducted the Quality-of-life assessment according to the Urbach scale questionnaires (scoring system 10-31, with 10 representing symptom free) which revealed mean ASQ score of 19.4[8]. Patients who were required reintervention had worse postoperative Eckardt and ASQ score with mean of 3.7 (P=0.53) and 22.7 (P=0.62) respectively. Twenty-one patients were over 16 years old and therefore too old for ongoing Paediatric care in our institute, of these 12/21 (57%) had a formal transition of care to adult services and had recurrent symptoms which required reintervention in 58% at some point. 9/21(43%) missed follow up after age of 16.

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Discussion

Esophageal achalasia (EA) is a rare diagnosis among the Paediatric age group, representing less than 5% of all achalasia cases and being extremely uncommon in children under 5 years old. This rarity contributes to diagnostic delays and a lack of standardized management strategies[9].

In our cohort, the predominant symptoms were regurgitation of undigested food, dysphagia, and malnutrition. Less typical symptoms included chest pain, heartburn, and respiratory complaints—findings that align with existing literature. The clinical presentation underscores the obstructive nature of the disorder and its significant nutritional and quality-of-life impact in children[10,11].

The diagnosis of EA relies on high-resolution manometry, which also guides treatment decisions based on the Chicago classification[12]. However, during the early study period, Paediatric manometry probes were unavailable, and manometry was performed in only 57% of patients. EA subtype was determined in 13 patients, with the majority being type II and one case of type III. The identification of subtypes is crucial, as type II typically demonstrates the best therapeutic response, whereas type III has the poorest outcomes[13].

The management of Paediatric EA is aimed at alleviating lower esophageal sphincter pressure rather than achieving a cure. Thus, all available treatments are considered palliative. Endoscopic Treatments, Botulinum toxin injection is discouraged as a first-line treatment per ISDE guidelines[14]. Balloon dilation remains an effective intervention, though 30–75% of children require repeat procedures[15,16]. Czubkowski observed better outcome of Heller myotomy when preceded by balloon dilatation and commented on its use as a bridge to improve dysphagia before myotomy[17], however others reported a negative effect on subsequent Heller myotomy[18]. In our cohort, 7% underwent balloon dilation as the primary treatment, yielding temporary relief but necessitating subsequent interventions. Surgical Treatments**,** the majority of our patients (93%) underwent Heller's myotomy with an antireflux procedure, transitioning from laparotomy to laparoscopy after 2009. Laparoscopic Heller's myotomy remains the standard approach in our institution. Although Peroral Endoscopic Myotomy (POEM) is increasingly used internationally, it is not currently available for Paediatric cases in Ireland.

In our study, the mean Eckardt score improved significantly from 6.9/12 preoperatively to 2.9/12 postoperatively ($p < 0.001$), indicating symptomatic relief. However, the mean ASQ quality-of-life score (19.4/31) suggests persistent symptoms post-intervention. Seventy-five percent of patients showed immediate and sustained postoperative improvement for an average of 17.4 months—comparable to findings by Goneidy et al[19]—yet 52% required further interventions, reflecting the non-curative nature of current therapies.

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These outcomes align with previous systematic reviews, such as Pacilli and Davenport, who reported an 85% short-term success rate for laparoscopic Heller's myotomy but highlighted the lack of long-term data[20]. Our findings reinforce that symptom recurrence is common, emphasizing the chronic, relapsing nature of EA.

A notable concern was the lack of formal transition to adult services in 43% of cases. This may reflect mild residual symptoms in some patients but nonetheless highlights the need for structured transition pathways. EA requires lifelong monitoring due to its chronicity and potential for relapse, making multidisciplinary continuity of care essential.

Our study is a retrospective single-center design, limited by the small sample size inherent to this rare condition, incomplete manometric data during early years, and variable follow-up durations. These factors restrict long-term outcome interpretation and generalizability.

Current management of Paediatric EA remains largely palliative. Future priorities include: Establishing long-term, multicenter registries for outcome tracking, expanding access to advanced modalities such as POEM, integrating standardized tools like ASQ into routine assessments to monitor disease burden and developing formal transition-of-care pathways to ensure continuity into adulthood.

In conclusion, childhood esophageal achalasia is a rare, chronic disorder requiring precise diagnosis, individualized treatment, and lifelong follow-up. While laparoscopic Heller myotomy offers effective short-term relief, symptom recurrence is common. The chronicity of EA necessitates ongoing care coordination and the exploration of more durable therapeutic approaches.

Declarations of Conflicts of Interest:
None declared.

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Appendix:

Table 1: *Summary of the patient's characteristics.*

patient's characteristics	
Mean age at diagnosis(range)-Years	11.7(1.4-15)
Gender	
Male	13(46%)
Female	15(54%)
Isolated EA	17
EA with co-morbidities	11
Down syndrome	3
Triple A syndrome	1
Raynaud's syndrome	1
Diabetus mellites	1
Scolioses	1
Esophageal atresia +TEF	1
Respiratory problem	3
Presenting Symptoms	
Regurgitation	28
Dysphagia	15
Chest pain	5
Failure to thrive	3
Weight loss	7
Investigation	
Contrast study	27(96%)
Endoscopy	26(93%)
High resolution-Manometry	16(57%)

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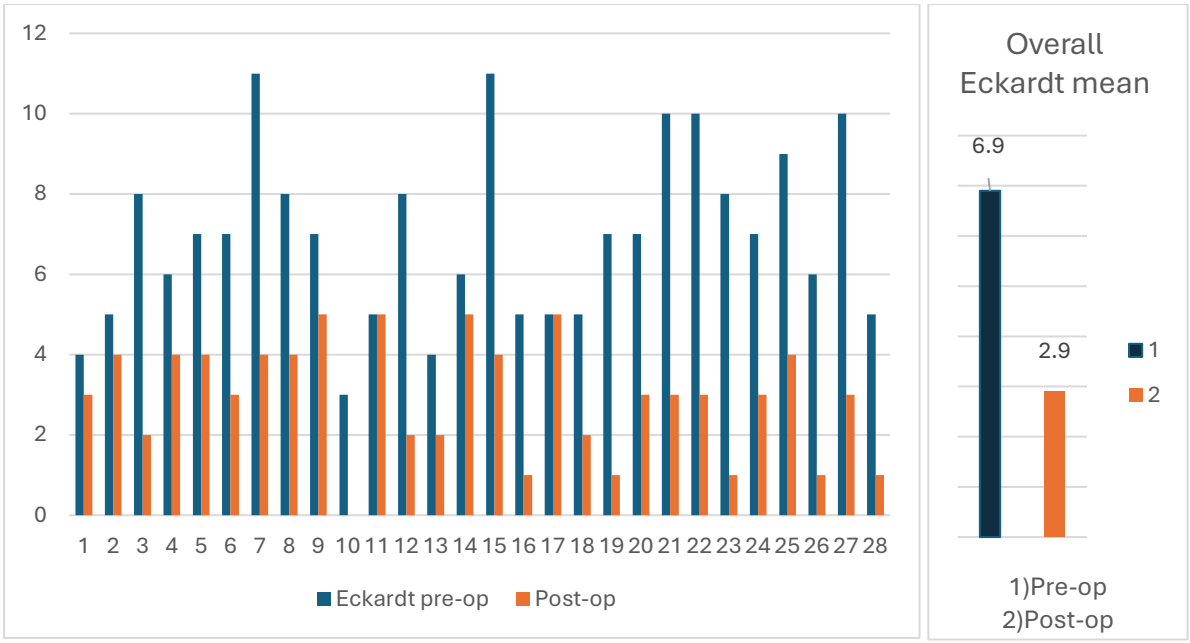

Fig 1: *Eckardt score of EA patients pre- and post-operative.*

Supplementary Data 1: *Esophgeal Achalasia severity: Eckardt Score*

Score for each sign/symptom

Sign/Symptom	0	1	2	3
Dysphagia	None	Occasional	Daily	Each meal
Chest pain	None	Occasional	Daily	Several times/day
Regurgitation	None	Occasional	Daily	Each meal
Weight loss(KG)	None	< 5	5-10	>10

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Supplementary Data 2: *Achalasia Score Questionnaire (ASQ). Quality of life assessment according to Urbach scale questionnaire.*

1. How much has achalasia limited the types of food you have been able to eat in the last month?
a. Not limited at all (I can eat and drink all the foods that I would like to.)**1**
b. Somewhat limited (I can eat and drink most of the foods that I would like to.)**2**
c. Moderately/Severely limited (I can eat and drink very few of the foods I would like to.)**3**

The following is a list of food types that may or may not cause you difficulty in swallowing. Please indicate which of the following types of foods you are able to swallow without experiencing any problems such as pain or food 'sticking' as it goes down. If you are not sure whether you can swallow a type of food without problems, please make you best guess.

2. Raw hard fruits and vegetables
a. Can swallow without problem.**1**
b. Can swallow, but with some difficulty.**2**
c. Can swallow with great difficulty or not at all.**3**

3. Rice
a. Can swallow without problem.**1**
b. Can swallow, but with some difficulty.**2**
c. Can swallow with great difficulty or not at all.**3**

4. Clear fluids (water, juice, coffee, tea)
a. Can swallow without problem.**1**
b. Can swallow, but with some difficulty.**2**
c. Can swallow with great difficulty or not at all.**3**

5. How often in the past month have you need to drink water while eating to deal with food caught in your esophagus?
a. Never/Rarely.**1**
b. Sometimes.**2**
c. Frequently/every time I eat.**3**

6. How often have you experienced pain when eating during the past month?
a. Never.**1**
b. Rarely.**2**
c. Sometimes.**3**
d. Frequently/every time I eat.**4**

7. During the past month, how much of a problem was heartburn (a burning pain behind the lower part of the chest) for you?
a. No problem.**1**

b. Mild problem.**2**
c. Moderate problem.**3**
d. Severe problem.**4**
e. Very severe problem.**5**

8. When you sit down to eat a meal, are you bothered by how long it takes you to finish eating?
a. No, I eat as quickly as I like.1
b. Yes, I am bothered by how long it takes me to eat.**2**

9. Has having achalasia limited your lifestyle?
a. No, it is not at all limiting (My daily activities have not changed.)**1**
b. Yes, it has limited my lifestyle (it has affected some areas, and I can no longer participate in all activities I want to do.)**2**

10. How much do you agree with the following statement about how satisfied you are with your health in regard to achalasia? I am satisfied with my health in regard to achalasia.
a. Strongly agree.**1**
b. Agree.**2**
c. Neither agree or disagree3
d. Disagree.**4**
e. Strongly disagree.**5**