REVIEW ARTICLE

Scoliosis after thoracotomy repair of esophageal atresia: a systematic review

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Abstract

Standard surgical repair of esophageal atresia/tracheoesophageal fistula (EA/TEF) is via a right posterolateral thoracotomy. A recognized complication is the later development of scoliosis. The prevalence and pathogenesis of secondary scoliosis are poorly understood. We, therefore, conducted a systematic review on this topic. All English language articles reporting incidence, outcomes and/or interventions for scoliosis in children after EA repair via thoracotomy were identified. Fourteen relevant articles published between 1969 and 2019 reporting 1338 children were included in the analysis. The aggregate prevalence of scoliosis among 937 children without congenital vertebral anomalies was 13%, but this figure varied widely between studies. Severity of scoliosis was documented in 181 children; eight children had a Cobb angle > 40° and 10 had undergone spinal surgery. The spinal curvature in affected individuals was dominantly or exclusively convex to the left. In conclusion, the reported prevalence of scoliosis varies widely but on average affects about one in eight children after open repair of EA/TEF. Most cases are mild and do not require intervention. It is currently uncertain whether secondary scoliosis is preventable by using meticulous thoracotomy techniques or thoracoscopic repair.
Level of evidence IV.

Keywords Tracheoesophageal fistula · Musculoskeletal · Thoracic surgery

Introduction

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) affects approximately 1:4000 live births [1]. Since 1941, the standard surgical repair has been via a right-sided posterolateral thoracotomy. A recognized complication of neonatal thoracotomy is the later development of secondary scoliosis. The incidence of secondary scoliosis may have declined in the modern era with the use of muscle-sparing thoracotomy techniques and avoidance of rib resections and pericostal sutures. Children with EA may also develop a primary scoliosis due to associated congenital vertebral and musculoskeletal malformations [2]. The prevalence and pathogenesis of secondary scoliosis are poorly understood.

This hinders our understanding of its prevention and the potential impact of thoracoscopic EA repair. We undertook a systematic review of the literature to investigate what is currently known about this topic.

Methods

A systematic review was conducted according to the PRISMA statement [3].

Identification of studies

A literature search was undertaken using MEDLINE, EMBASE and the first ten pages of Google Scholar using the following keywords: "esophageal atresia" and "scoliosis". The search was completed on 31st October 2019. Reference lists of all relevant articles were searched for additional studies.

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Inclusion/exclusion criteria

All English language studies reporting on incidence, outcomes or interventions for scoliosis in children born with EA and undergoing repair via thoracotomy were included. The following studies were excluded: case reports; those reporting duplicate or overlapping patient cohorts; review articles not providing any novel patient data; studies including patients undergoing thoracotomy for other conditions; and studies with unknown scoliosis outcomes.

Study selection and data extraction

Two reviewers (PM, GT) screened the abstracts of all potentially relevant studies. In case of disagreement, a third independent reviewer was consulted (MS). After screening, relevant full-text articles were obtained and reviewed independently by all three authors using a standardized data-extraction template.

Outcomes

The incidence, severity, laterality, and risk factors for scoliosis were analyzed along with the need for intervention.

Results

Analysis

The initial search yielded a total of 197 results from which 34 articles were selected for full-text assessment (Fig. 1). After exclusions, 14 articles were included in the final analysis. Two of these studies included a small proportion of patients who had undergone thoracoscopic EA repair [4, 5], but since more than 93% of each cohort had been treated by thoracotomy these studies were included.

All studies were retrospective analyses of an institutional cohort; one was a case–control study [6]. Studies were published between 1969 and 2019. There was considerable
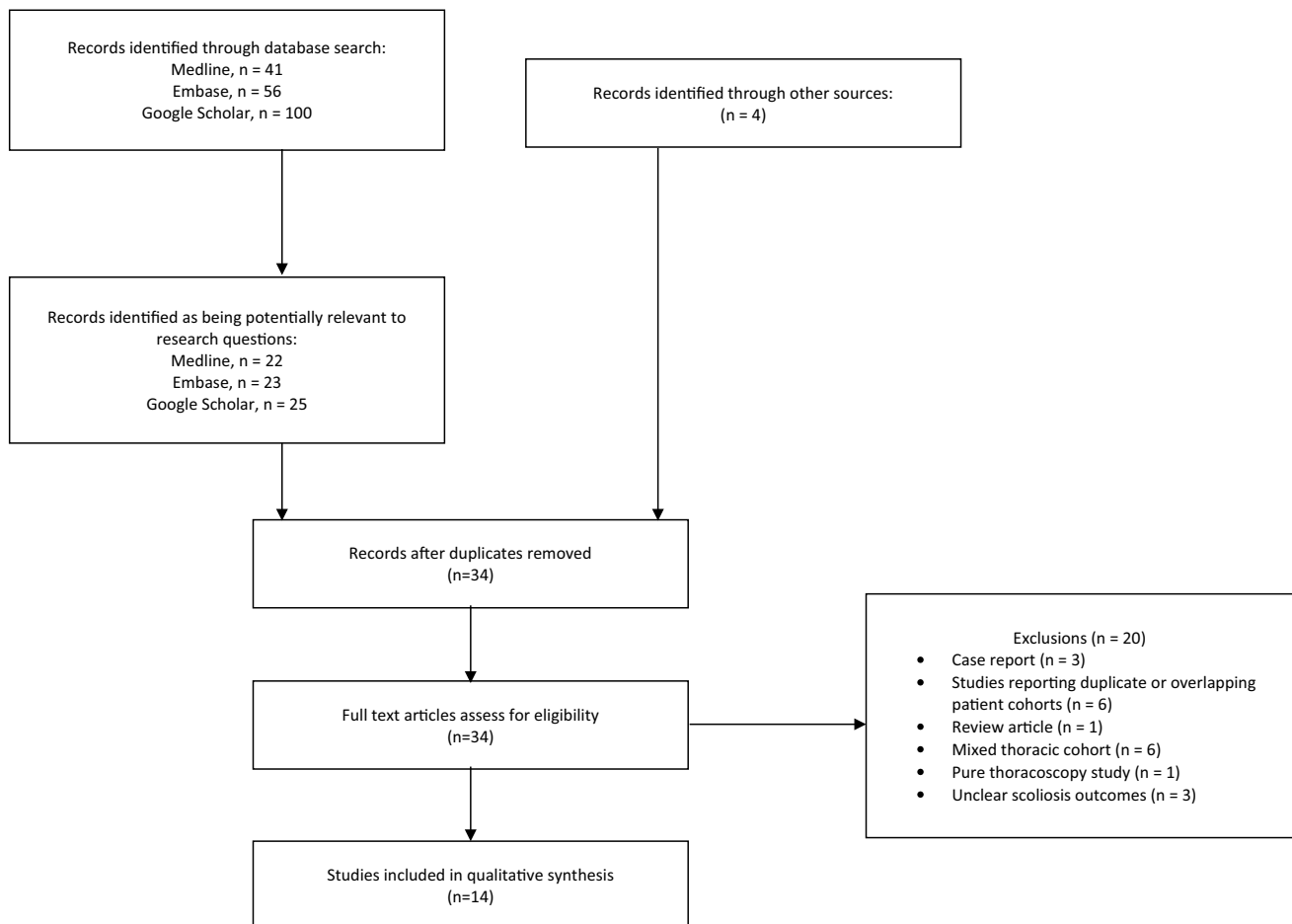

Fig. 1 PRISMA flow diagram

heterogeneity between studies in relation to surgical era, thoracotomy technique, age at assessment, and exclusion of congenital vertebral or other relevant musculoskeletal anomalies, making pooled analysis of data inappropriate. In addition, there was a response bias in almost all studies. For example, 19 of 58 (33%) survivors were evaluated by Freeman and Walkden [7], 101 of 235 (43%) potentially eligible survivors by Sistonen et al. [6], 30 of 51 (59%) by Koziarkiewicz et al. [8] and 322 of 397 (81%) patients by Bastard et al. [5]. The potential contribution of repeated thoracotomy or additional thoracic surgery for congenital cardiac disease was not stated or unknown.

Outcomes

Collectively, the 14 studies comprised 1338 children with 268 (20%) reported as having scoliosis after thoracotomy for EA repair (Table 1) [4, 5, 7–18]. The incidence of scoliosis varied greatly between 3 and 67% and this was evident in both older and recent cohorts; a recent national French cohort recorded a rate of 4% while a Canadian study reported a rate of 50% [4, 5].

Seven studies [4, 12, 14–18] reported on the severity of scoliosis assessed by the Cobb angle [19]. Assessments were conducted with variable rigor, the best methodology being adopted by Sistonen et al. [6] in which spinal radiographs were graded independently by two orthopedic surgeons. The majority of affected patients had mild scoliosis not requiring intervention, with a Cobb angle of less than 20°. Of the seven studies reporting Cobb angles in 181 children with scoliosis after open EA/TEF repair, eight children had a Cobb angle exceeding 40° at the time of reporting and 10 had undergone spinal surgery. However, it should be noted that the length of follow-up in individual studies was very variable and several authors emphasized that scoliosis often only becomes apparent or clinically relevant in teenage years [6, 17].

Ten studies reported on the incidence of associated congenital vertebral anomalies [4, 5, 7, 8, 11–13, 15, 16, 18] and a further two excluded these patients from their analysis [9, 17]. The proportion of children with EA/TEF and congenital vertebral anomalies varied widely between 4 and 45% but in only one study was this figure greater than 20% [6, 12]; it is notable that this was a study in adults aged between 21 and 57 years and the authors commented that only 11% of these anomalies were diagnosed in childhood. However, the majority of these vertebral anomalies were in the cervical spine and quite possibly only detected because spinal radiographs were taken for the study. Two studies reported scoliosis outcomes separately for children with congenital vertebral anomalies [4, 15]. In a Canadian study, all eight children with an associated vertebral anomaly developed scoliosis and although the authors stated that none

had progressed to requiring surgical intervention, they were only aged 3–13 years at assessment [4]. In the Melbourne series of patients with EA/TEF, many of whom were followed into adulthood, 53 (19%) had an associated congenital vertebral anomaly and one-third subsequently developed scoliosis [15].

After excluding patients with congenital vertebral anomalies pooled estimates from seven studies [4, 5, 9, 11, 15–18] showed that 124 of 937 (13%) infants developed scoliosis after open EA/TEF repair. This figure varied widely between different cohorts in studies published before 1990 (10–50% prevalence) and studies published after 2010 (0–46% prevalence).

Five studies specifically reported on the direction of the thoracic curve in patients with scoliosis after open EA/TEF repair [4, 15–18]. Four of these described the spinal curvature as dominantly or exclusively convex to the left i.e. to the side opposite a right posterolateral thoracotomy [15–18]. In contrast, Soliman et al. reported that 33 of 46 patients (72%) with a thoracic scoliosis had a curve convex to the right i.e. convex towards the side of the thoracotomy (89% of their cohort had undergone a right-sided thoracotomy) [4]. The reasons for this discrepancy are unclear but the methodology used in the Canadian study was not as robust as that used by Sistonen et al. who reported that the main scoliotic curve was upper thoracic in 31 (55%) patients and was concave towards the side of the thoracotomy [6, 12].

Discussion

In countries with well-developed health care systems, the overall survival rate of children born with EA ± TEF exceeds 90%. In the absence of associated congenital heart disorders, major chromosomal anomalies or extreme prematurity this figure is close to 100% [20]. Surgical techniques of open repair have evolved over time with more widespread adoption of muscle-sparing incisions [21], better esophageal anastomotic techniques, and avoidance of tight pericostal sutures in chest wall closure. Nevertheless, an open thoracotomy may be complicated by significant musculoskeletal and cosmetic deformities that may not become apparent until the child is much older [13].

Scoliosis, an abnormal lateral curvature of the spine (greater than 10 degrees), is a complex three-dimensional rotational deformity. 'Idiopathic' scoliosis affects 2–3% of the general population, is more common in adolescent girls and more often convex to the right [22–24]. Whilst a congenital predisposition to idiopathic scoliosis among children with EA/TEF cannot be ruled out this is unlikely because scoliosis associated with EA/TEF shows no sex bias, is more often convex to the left and often first becomes evident before puberty [24]. Our systematic review indicates a

Table 1 Published studies of scoliosis after EA/TEF repair

Study (era)	No. assessed with repaired EA	Age at review (y)	Scoliosis	Congenital vertebral anomalies	Severity (Cobb angle)	Convexity to OPPOSITE side of thoracotomy (to left after right thoracotomy)	Surgical intervention
Soliman [4] (2005–2015) Canada	106 (82 EA+TEF)	7 (median) (range 5–14)	53 (50%); 46 in thoracic region 45/98 (46%) after excluding congenital vertebral anomalies	8/106 (8%); all developed scoliosis	40 curve 10–20° 2 curve 20–30° 7 curve 30–45° 4 curve >45°	13/46 (28%)	4/53 surgical stabilisation None with congenital vertebral anomaly
Bastard [5] (2008–2010) France	322 (285 EA+TEF)	2.8 (mean)	12/297 (4%) after excluding congenital thoracic malformations	25/322 (8%) thoracic malformations 14/322 (4%) thoracic hemivertebra	Not stated	Not stated	Not stated
Wei [9] (1997–2011) Canada	52 (46 EA+TEF)	8 (median)	4 (8%)	Excluded	"Subclinical"	Not stated	0
Okuyama [10] (1970–2000) Japan	69 (63 EA+TEF)	19 (median)	21/62 (34%) with EA+TEF 4/6 isolated EA	Unknown	Not stated	Not stated	6/69 surgical stabilisation
Koziarkiewicz [8] (1990–2005) Poland	30 (24 EA+TEF)	13.7 (mean)	20/30 (67%)	6/30 (20%)	Not stated	Not stated	Not stated
Burford [11] (1993–2008) USA	72 (All EA+TEF)	1–16 (mean 11y for 2 with scoliosis)	2/72 (3%) 0 after excluding congenital vertebral anomalies	3/72 (4%)	"Mild"	Not stated	0
Sistonen [12][a] (1947–1985) Finland	101 (91 EA+TEF)	36 (median) (range 21–57)	56 (56%)	45/101 (45%): 38 cervical, 15 thoracic; 5 lumbar	45 curve 10–20° 10 curve 20–45° 1 curve >45°	"Most common"	0
Mortell [13] (1997–2007) USA	46	6 (median)	3/46 (7%)	6/92 (7%)	Not stated	Not stated	Not stated
Somppi [14] (1963–1993) Finland	42	12.6 (mean)	9/42 (21%)	Unknown	5 to 37°	Not stated	Not stated
Chetcuti [15] (1948–1985) Australia	285	1–38	48/285 (17%) 30/232 (13%) after excluding congenital vertebral anomalies 16/173 (9%) after single thoracotomy and no vertebral anomaly	53/285 (19%); 18 of whom developed scoliosis	Only stated for those without congenital vertebral anomalies (n=30): 10 minimal 17 mild (15–20°) 2 moderate (20–25°) 1 curve severe (≥40°)	20/30 without congenital vertebral anomalies	1/232 patients without congenital vertebral anomalies 8/53 with congenital vertebral anomalies

Table 1 (continued)

Study (era)	No. assessed with repaired EA	Age at review (y)	Scoliosis	Congenital vertebral anomalies	Severity (Cobb angle)	Convexity to OPPO-SITE side of thoracotomy (to left after right thoracotomy)	Surgical intervention
Jaureguizar [16] (1965–1981) Spain	89 (All EA+TEF)	6.3 (mean)	16/89 (18%)	6/89 (7%); excluded from study	7 curve >20°	16/16	0/16 surgical stabilisation 0/16 bracing
Gilsanz [17] (1966–1980) USA	82	8.5 (mean)	8/82 (10%)	Excluded	1 with 30° scoliosis by 2y; Scoliosis >20° in another 7 but only appeared after 11y	8/8	4/8 surgical stabilisation
Durning [18] (1960–1970) USA	18	11.8 (mean)	9 (50%)	5/39 (13%); excluded from study	4 curve 10–20° 3 curve 20–40° 2 curve >40°	8/9	1/9 surgical stabilisation 3/9 bracing
Freeman [7] (1955–1965) UK	24	3–14	3 (13%)	1/19 (5%)	2 curves "mild" 1 curve "moderate"	Not stated	Not stated

EA esophageal atresia, *TEF* tracheoesophageal fistula

[a] Additional data from Sistonen et al. [6]

wide variation in the reported prevalence of scoliosis after open EA/TEF repair with figures ranging from 3 to 67%. This variance may relate to multiple factors: different definitions of scoliosis and methods of ascertainment; technical differences in thoracotomy and chest wall closure; variable proportions of patients lost to follow-up; variable duration of follow-up; and inconsistent exclusion of patients with associated vertebral anomalies.

After excluding infants with congenital musculoskeletal anomalies our systematic review indicates that approximately 13% of infants develop scoliosis after open EA/TEF repair, although this figure varies widely between studies. Most cases are relatively mild and do not require surgical correction. Numerous risk factors for the development of scoliosis have been suggested including repeated thoracotomy [13, 15], rib resection [15], acquired rib fusion from tight intercostal closure [4, 6, 15, 17], division of the serratus anterior muscle [9], and anastomotic leak leading to empyema and pleural scarring [9, 15, 17]. Whilst rib resections and vertical parascapular incisions are now obsolete, we could not demonstrate a clear era effect in our analysis. Thus, the prevalence of scoliosis among patients without a congenital vertebral anomaly varied widely in studies published before 1990 [15–18] and in those published after 2000 [4, 5, 9, 11]. However, with one exception [4], the frequency of scoliosis tended to be lower in more recent cohorts.

Thoracotomy in infancy for other indications such as congenital cardiac disease [25], ligation of patent ductus arteriosus or repair of coarctation of the aorta [26] and congenital lung malformations [27] may also be complicated by later scoliosis. Roclawski et al. [26] evaluated 126 patients after repair of coarctation of the aorta or patent ductus arteriosus during childhood; 83 underwent repair through a left posterolateral thoracotomy and 43 were managed non-operatively using interventional radiologic techniques. The prevalence of scoliosis was 59% in the thoracotomy group after a mean follow-up of 15 years compared to 19% in the non-operative group after a mean of 9 years. Most curves were mild but six individuals in the thoracotomy group had curves ≥ 30°. After thoracotomy, most curves were thoracic with an almost equal distribution between right and left convex curves as compared to curves convex to the right in the non-operative patients. Kaito et al. [25] analyzed the chest radiographs of 483 children who had undergone surgery via thoracotomy and/or sternotomy for congenital cardiac disorders in the first year of life. After a mean follow-up of 14 years, one-third had a mild scoliosis but 5% had curves ≥ 30°. However, as the majority of children who had had a thoracotomy had also undergone sternotomy the role of thoracotomy alone on the development of scoliosis is difficult to define. Since infants undergoing median sternotomy [25, 28] or repair of congenital diaphragmatic hernia [29] are also at increased risk of scoliosis, even in the absence of congenital vertebral

anomalies, the pathogenesis of secondary scoliosis is clearly complex. Mechanical injury to the costovertebral joints after sternotomy or thoracotomy may be contributory [25], as may damage to consecutive intercostal nerves [30]. Both these mechanisms are supported by experiments in rabbits [31]. With the exception of one study [4], the direction of spinal curvature in post-thoracotomy scoliosis following EA repair is dominantly or exclusively convex to the left.

Uncomplicated thoracoscopic repair of EA/TEF might reduce the risk of scoliosis and other potential chest wall deformities such as winging of the scapula. Current published results of thoracoscopic EA/TEF repair report no "clinically significant scoliosis" [32, 33], but studies of larger numbers of patients assessed after longer follow-up periods are required before this can be accepted as a definitive long-term outcome.

Our systematic review is limited by the heterogeneity of the data. However, according to the available published literature, scoliosis develops in about one in eight children without congenital vertebral anomalies after thoracotomy repair of EA/TEF. Multiple factors are implicated in the pathogenesis. Most cases are mild and do not require surgical intervention. It is as yet uncertain whether secondary scoliosis is avoidable by meticulous thoracotomy techniques or thoracoscopic repair.

Compliance with ethical standards

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