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Natural History of Spine in Patients with Esophageal Atresia -A Long-Term Population-Based Follow-up Study

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Authors Disclosure

Information

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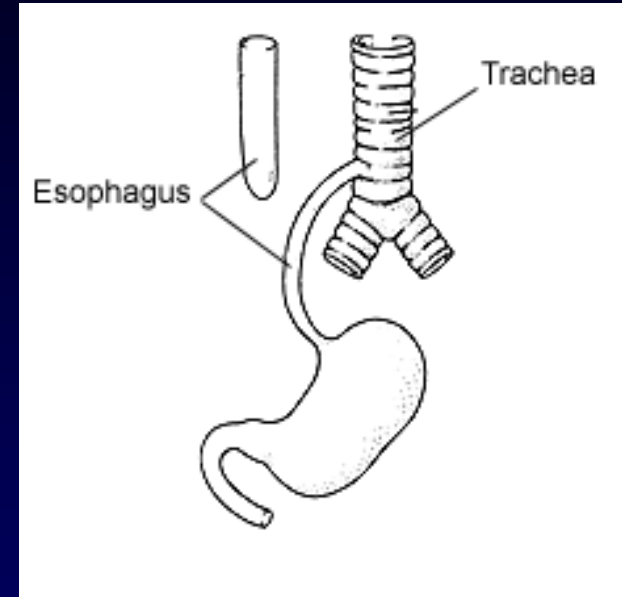
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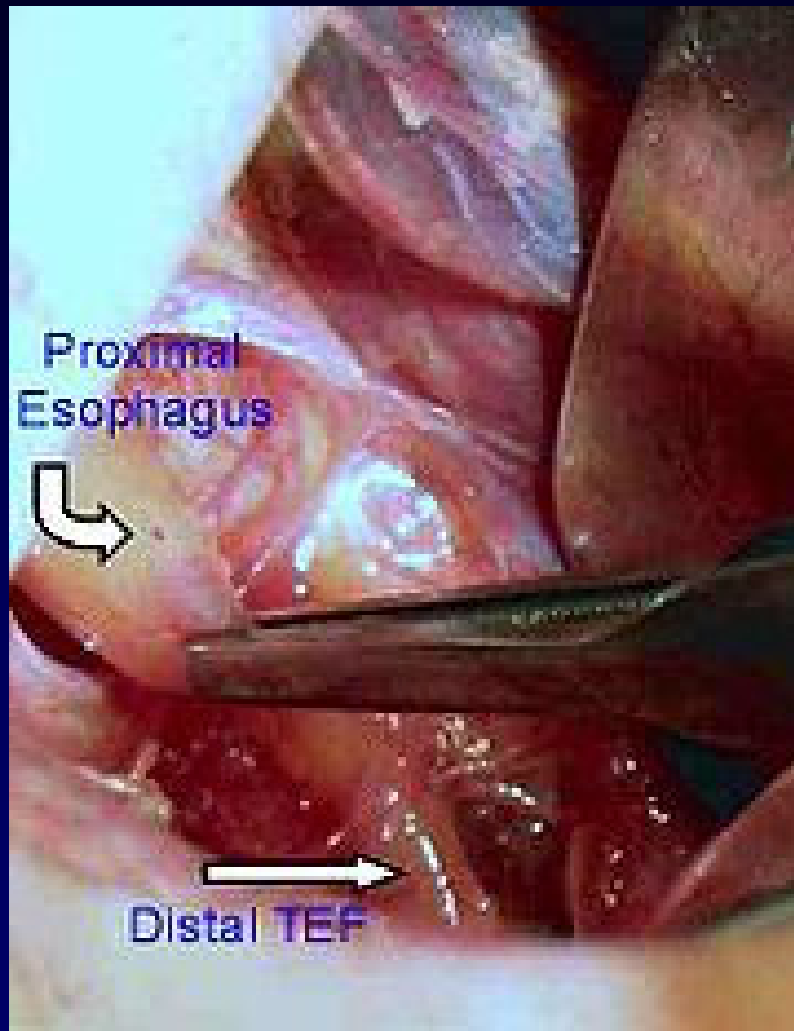
Introduction

- Vertebral column malformations typically associate esophageal atresia (EA) (Lawhon et al. JBJS 1986)
- VATER association
 - Vertebra, Anus, Trachea, Esophagus, Renis/Radius anomalies
- Thoracotomy during neonatal period increases further risk of scoliosis (Durning et al. JBJS 1980)
- One-fourth of patients with EA have vertebral anomalies (Keckler Ped Surg Int 2007)
- Natural history of scoliosis and vertebral anomalies in patients with EA unknown



Esophageal atresia with tracheoesophageal fistula

Esophageal atresia repair



Repair during neonatal period

Right thoracotomy

Tracheoesophageal fistula seclusion

Primary Esophago-Esophagostomy

Methods

- 588 patients operated for EA in Helsinki Children's Hospital between 1949 and 1985 were identified.
- 296 were alive and contacted in 2005.
- First 100 returning signed informed consent participated (Table 1).
- All patients had follow-up to adulthood, mean follow-up 36 yrs (21-57 yrs)

Study design



- Retrospective baseline data collection.
- All 100 patients underwent:
 - Physical examination of the spine
 - A standing PA spine radiograph
 - AP and lateral radiograph of c-spine
 - Questionnaire about skeletal symptoms
- Control population of 855 school children, if rib hump > 6 deg \rightarrow Scoliosis radiograph (Nissinen 1993)

Patients

	Participants (n=100)	Non-participants (n=161)
Age (yrs)	36 (21-57)	37 (21-57)
Males (%)	57 (57%)	84 (62%)
Distal TE fistula	91 (91%)	120 (89%)
VACTERL		
Primarily	5 (5%)	8 (5%)
Currently	23 (23%)	

Clinical findings



- 13 (13%) back or neck pain often at rest
- 54 (54%) showed rib hump $\geq 6^\circ$
- 15 (15%) asymmetry of anterior thoracic wall

Vertebral anomalies



- 45 (45%) at least one vertebral anomaly.
- Cervical spine block vertebrae most common anomaly
- None with spine surgery

Results

Vertebral anomalies

Anomaly	Number (%)
Failure of segmentation	
Fused vertebrae	43 (43%)
Unilateral bar	4 (4%)
Failure of formation	
Wedge vertebrae	22 (22%)
Butterfly	8 (8%)
Hemivertebra	3 (3%)

Results

Risk of scoliosis

	Patients	Controls	OR (95% CI)
>10 degrees	56 (56%)	79 (9.2%)	13.2 (8.3-21)
≥25 degrees	11 (11%)	6 (2.4%)	37.8 (14-106)
≥45 degrees	1 (1%)	0	

Risk factors for Scoliosis

Multiple Logistic Regression Model

Variable	OR (95% CI)	P value
Gender	1.2 (0.6-2.9)	0.60
Heart surgery	2.0 (0.4-11)	0.40
Rib fusions	3.6 (0.7-19)	0.13
Any additional anomaly	2.1 (0.9-4.8)	0.07

Conclusions



- Risk of scoliosis 13-fold after repair of EA.
- Nearly half of patients present with vertebral anomalies, most commonly in cervical spine.
- Further assessment of cervical spine with full extension-flexion radiographs indicated to rule out instability.

Conclusions cont'd

- Associated anomalies most significant risk factor for scoliosis.
- Cervical vertebrae and esophagus anatomically adjacent structures, a local developmental defect in this area may underpin the synchronous occurrence of these anomalies.
- Spinal surgery rarely indicated, but due to high incidence routine screening of spine necessary



Thank you

References

Lawhon SM, MacEwan GD, Bunnell WP. J Bone Joint Surg 1986;68:424-9.

Keckler SJ, Peter SD, Valusek PA, et al. Ped Surg Int 2007;23:309-13.