Spinal deformity in patients born with oesophageal atresia and tracheo-oesophageal fistula

P CHETCUTI, D R V DICKENS,* AND P D PHELAN

*Department of Paediatrics, University of Melbourne, and *Department of Orthopaedic Surgery, Royal Children's Hospital, Melbourne, Australia*

SUMMARY Spinal deformity was present in 58 (19%) of 302 patients born with oesophageal atresia and fistula when examined at review. This was present in 24 (47%) of 51 patients with vertebral anomalies and 34 (14%) of 251 patients with normal vertebrae. Scoliosis was present in 21 patients with vertebral anomalies, torticollis in two, and lordosis in one. Two thirds of the patients with congenital scoliosis have had or are likely to require operation, compared with five patients with non-congenital scoliosis. Scoliosis associated with mixed vertebral anomalies in the lower thoracic spine had the worst prognosis. The medical records of a further 64 patients who had survived operation but who could not be traced were reviewed, and indicated that four had had congenital vertebral anomalies but none had a spinal deformity.

We recommend early detection of vertebral anomalies in this group and careful follow up of patients with these abnormalities.

Morbidity after repair of oesophageal atresia and fistula is not limited to respiratory[1] and surgical complications.[2] Spinal deformity has been reported in later childhood,[3–5] and may be related to associated congenital vertebral anomalies,[6–8] the thoracotomy,[3 4] and chronic pleural infection.[3] Previous reports of scoliosis in patients born with oesophageal atresia and tracheo-oesophageal fistula have been limited to small numbers and have excluded patients with congenital vertebral abnormalities.

The present report is a clinical and radiological review of 366 patients born with oesophageal atresia and tracheo-oesophageal fistula.

Patients and methods

Five hundred and thirty eight patients with oesophageal atresia and tracheo-oesophageal fistula have been treated at the Royal Children's Hospital, Melbourne, between 1948 when the first successful repair was carried out, and the end of 1985. Between August 1986 and April 1987, 302 of the 366 survivors were interviewed and examined as part of a long term follow up study. The patients were aged 1 to 37 years with a mean age of 18 years, and 59% were men or boys. Radiographs from the time of initial diagnosis as well as any performed subse-

quently were available for assessment in 333 patients, and radiological reports alone in the remaining 33. Each patient was examined for evidence of scoliosis, and their previous radiographs reviewed. Ten children and adolescents with scoliosis not previously recognised had new spinal radiographs. Twelve adults with minimal scoliosis did not have new radiographs as they had no symptoms referrable to the back. The Cobb angle was measured.[9] There were no data in the medical records of the 64 patients who were not examined to suggest that they had had scoliosis or other spinal deformity.

Vertebral anomalies were classified as: segmentation defects consisting of unilateral unsegmented bars and block vertebrae; formation defects consisting of complete (hemivertebrae) or partial defects (wedge vertebrae); or mixed defects consisting of unilateral failure of segmentation with contralateral hemivertebrae at the same level. Partial and complete sacral agenesis and absence of the coccyx were included in formation defects. Numerical rib anomalies were documented.

The scoliotic deformity was subdivided into an upper thoracic curve if the apex lay between the second and sixth thoracic vertebrae; a lower thoracic curve if it lay between the seventh and 11th thoracic vertebrae; a thoracolumbar curve if the apex was at

the 12th thoracic or first lumbar vertebrae; a lumbar if it lay between the second and fourth lumbar vertebrae, and a lumbosacral if the apex was at the fifth lumbar vertebra.

Total lung capacity and vital capacity were measured in a pressure compensated plethysmograph (Jaeger Bodyscreen) in patients over the age of 6 who were able to cooperate. The best of a minimum of three efforts in the sitting position was recorded. Values were expressed as percent predicted for age, height and sex, based on the normal values of Zapletel *et al*[10] and Knudson *et al.*[11][12]

The unpaired Student's *t* test was used to analyse lung function and the χ^2 test to determine significance of differences between congenital vertebral abnormalities and other congenital abnormalities.

Results

Sixty seven congenital vertebral abnormalities were documented radiologically in 55 patients. They were equally distributed among the different types of oesophageal atresia but only one out of 26 patients with an isolated tracheo-oesophageal fistula had a vertebral anomaly. The types of anomalies present are given in table 1. Eleven patients had a combination of defects at different vertebral sites. Patients with anorectal and rectal anomalies were more likely to have vertebral anomalies ($p<0.05$). Numerical rib abnormalities, present in 81 patients, were more common in those with vertebral abnormalities.

Spinal deformities were clinically apparent in 24 (47%) of the 51 patients with vertebral abnormalities who attended for review: scoliosis in 21 (confirmed radiologically), torticollis in two, and a lordosis in one. The types and distribution of anomalies in the 21 patients with scoliosis are given in table 2.

Table 1   *Types and distribution of congenital vertebral anomalies in 55 patients*

	Formation defects	Segmentation defects	Mixed defects
Cervical	0	3	0
Cervicothoracic	2	3	3
Upper thoracic	8	2	4
Lower thoracic	8	0	6
Thoracolumbar	1	3	2
Lumbar	1	2	1
Lumbosacral	6	3	0
Sacral	8	0	0
Coccygeal	1	0	0
Total	35	16	16

Table 2   *Sites of congenital scoliosis in patients with vertebral anomalies*

	Formation defects	Segmentation defects	Mixed defects
Total with scoliosis	10	4	7
Upper thoracic	1	2	1
Lower thoracic	5	0	5
Thoracolumbar	1	2	1
Lumbar	0	0	0
Lumbosacral	3	0	0

Surgical stabilisation of the spine was performed in eight patients: seven during adolescence and one in adult life. Mixed deformities in the lower thoracic spine were present in three of these, formation defects in the lower thoracic spine in two, a segmentation defect in the upper thoracic spine in one, a mixed defect in the thoracolumbar spine in one, and a formation defect in the thoracolumbar spine in one patient. Operation was not carried out in one adult patient with a segmentation defect and a 45° thoracolumbar curve because he was also mentally retarded. Five preadolescent children had significant curves (>20°); lumbosacral formation defects and curves of 20° and 27° were present in two, lower thoracic mixed defects and curves of 20° in two, and a segmentation defect and a curve of 26° in one patient.

A mixed defect and a segmentation defect were found in the cervicothoracic spine of the two patients with torticollis. The patient with the segmentation defect had a severe torticollis. A lumbosacral spondylolisthesis was present in one patient who presented with a lumbar lordosis and had had it operated on.

A thoracic scoliosis was present in 34 patients (14%) with normal vertebrae. An upper thoracic curve was present in 32 and a lower thoracic curve in two. Surgical stabilisation of the spine was done during adolescence in one patient with an upper thoracic curve. Curves of 20° and 25° were seen on the radiographs of two preadolescent children, and curves of between 15 and 20° in 17 adolescent and adult patients. A mild scoliosis was present on clinical examination of 12 other patients. Operation was carried out during adolescence in one girl with a lower thoracic curve and the other, a preadolescent girl, had a curve of 22°. Scoliosis at other sites was not found in patients with normal vertebrae.

Lung function is summarised in table 3. Patients with scoliosis had a reduction in total lung capacity and vital capacity compared with those with a normal spine ($p<0.001$). Patients with congenital

Table 3 *Lung function results in 202 patients with repaired oesophageal atresia and tracheo-oesophageal fistula. Figures are expressed as mean % plus one standard deviation of predicted normal based on height*

	No scoliosis	All scoliosis	Congenital scoliosis	Non-congenital scoliosis
Total lung capacity	98·3+13·1	89·5+16·4	86·3+16·2	91·8+16·5
Vital capacity	89·5+16·4	75·8+19·2	74·3+21·4	76·8+17·7

scoliosis had a reduction of total lung capacity and vital capacity compared with patients with non-congenital scoliosis, but the difference was not significant.

Discussion

Vertebral anomalies and spinal deformity in patients born with oesophageal atresia and tracheo-oesophageal fistula are common. Congenital vertebral anomalies were present in 15% of patients in the present study, compared with an incidence of 2 to 45% in previous reports.[6 7 13 14] Those were based on small numbers or did not include specific radiological review. Our incidence is a minimal one as in 10% of patients we had only radiological reports and in some radiographs the whole spine was not shown. As previously reported, vertebral abnormalities were less common with isolated tracheo-oesophageal fistula, which is true of other anomalies as well.[1 15] The association of vertebral, renal, and anorectal anomalies is already well recognised and emphasises the importance of looking for latent renal abnormalities in patients with vertebral abnormalities.[16]

Vertebral anomalies cause an imbalance in the longitudinal growth of the spine caused by a deficiency either in the number of end plates, or in their rate of growth on one side of the spine.[17] The severity of the lateral curve that develops is proportional to the degree of the imbalance of growth. The formation defects were the most common type of anomaly causing scoliosis, and the lower thoracic spine the most common site. Two thirds of our patients with congenital scoliosis have had or are likely to require operation, compared with half quoted for congenital sciolosis.[18] Mixed defects in the lower thoracic spine seem to have the worst prognosis.[18 19]

Lumbosacral curves (as present in two of our patients) are potentially serious as there is no room below the vertebral defect for natural compensation to occur. This deformity is particularly difficult to correct.[20]

The prognosis for patients with scoliosis in the absence of congenital abnormalities is usually good. Operation was necessary, however, in two patients,

and three children had pronounced curves that may progress. The aetiology probably has several components, but rib resection during thoracotomy,[21 22] scar formation from the incision itself,[23] and chronic pleural infection with rib fusion[5 24] may all contribute. Scoliosis is commoner in patients who have had several thoracotomies.[4] One patient requiring operation and one of the three with pronounced curves, however, had lower thoracic scoliosis of the type typical of idiopathic scoliosis. Given that the prevalence of scoliosis with curves greater than 20° at age 14 is 5/1000,[25] finding two in this group of 302 patients is not surprising.

We confirmed that patients with scoliosis had an appreciable reduction in lung volume. While anthropometric techniques have been standardised for normal children, this is not the case for patients with scoliosis. Clearly height becomes progressively more inappropriate as the severity of scoliosis increases and its use may underestimate the degree of abnormality. Many scoliosis clinics are now using the measurement of arm span to normalise lung function, but adequate normal data for these measurements in healthy children were not available at the time of the study.

Congenital and non-congenital spinal curves are common in patients born with oesophageal atresia and fistula. The thoracotomy may further contribute to the progression of the congenital scoliosis. Routine screening of the spine at birth for congenital anomalies and careful follow up if they are found are recommended. Some anomalies are associated with a poor prognosis and early treatment is recommended.[26] It is far better to prevent an increase in the deformity than to correct it once it has become severe.

References

1 Dudley NE, Phelan PD. Respiratory complications in long term survivors of oesophageal atresia. *Arch Dis Child* 1976;**51**: 279–82.
2 Myers NA. Oesophageal atresia and/or tracheo-oesophageal fistula. A study of mortality. In: Rickham PP, Hecker WC, Prevot S, eds. *Causes of postoperative death in children.* Baltimore: Urban and Schwarzenberg, 1979:141–65.
3 Durning RP, Scoles PV, Fox OD. Scoliosis after thoracotomy in tracheo-oesophageal fistula patients. *J Bone Joint Surg (Am)* 1980;**62**:1156–8.

4 Chetcuti P, Myers NA, Phelan PD, Beasley SW, Dickens DRV. Chest wall deformity in patients with repaired oesophageal atresia. *J Pediatr Surg* 1989;**24**:244–7.

5 Gilsanz U, Boechat IM, Bimberg FA, King JD. Scoliosis after thoracotomy for oesophageal atresia. *AJR* 1983;**141**:457–60.

6 Louhimo I, Lindahl H. Oesophageal atresia: primary results of 500 consecutively treated patients. *J Pediatr Surg* 1983;**18**:217–29.

7 Holder TM, Cloud DT, Lewis JE, Pilling GP. Oesophageal atresia and tracheo-oesophageal fistula: a survey of its members by the surgical section of the American Academy of Pediatrics. *Pediatrics* 1964;**34**:542–9.

8 Bond-Taylor W, Storer F, Atwell JD. Vertebral anomalies associated with esophageal atresia and tracheo-esophageal fistula with reference to the initial operative mortality. *J Pediatr Surg* 1973;**8**:9–13.

9 Cobb JR. Outline for the study of scoliosis. In: Arbor A, Edwards JW, eds. *Instructional course lectures.* American Academy of Orthopaedic Surgeons, 1948;**5**:261–75.

10 Zapletal A, Paul T, Samanell M. Die Bedeutung leutiger methoden der lungen funklion zur Feststelling einer obstruktion der atemarege bei Kinder und Jugendlichen. *Z Erkr Atmungsorgane* 1977;**149**:343–71.

11 Knudson RJ, Slatin RL, Lebowitz MD. The maximal expiratory flow volume curve. Normal standards, variability and effects of age. *Am Rev Respir Dis* 1976;**113**:587–600.

12 Knudson RJ, Lebowitz MD, Holberg CJ, Burrows B. Changes in the normal maximal expiratory flow-volume curves with growth and ageing. *Am Rev Respir Dis* 1982;**127**:125–34.

13 Swenson O, Lipman R, Trisher JH, Deluca FG. Repair and complications of oesophageal atresia and tracheo-esophageal fistula. *N Engl J Med* 1962;**267**:960–3.

14 Martin LW. Management of oesophageal anomalies. *Pediatrics* 1965;**36**:342–50.

15 Schneider KM, Becher JM. The 'H-type' tracheo-oesophageal fistula in infants and children. *Surgery* 1962;**51**:677–86.

16 MacEwan G, Winter R, Hardy J. Evaluation of kidney anomalies in congenital scoliosis. *J Bone Joint Surg (Am)* 1972;**54**:1451–4.

17 Bick EM, Lopel JW. Longitudinal growth of the human vertebra. A contribution to human osteogeney. *J Bone Joint Surg (Am)* 1950;**32**:803–14.

18 Winter RB, Moe JH, Gilers VE. Congenital scoliosis. A study of 234 patients treated and untreated. Part 1: Natural history. *J Bone Joint Surg (Am)* 1968;**50**:1–15.

19 McMaster MJ, Ohtsuka K. The natural history of congenital scoliosis. A study of two hundred and fifty-one patients. *J Bone Joint Surg (Am)* 1982;**64**:1128–47.

20 Winter RB. *Congenital deformities of the spine.* New York: Thieme-Stratton, 1983.

21 Stauffer GS, Mankin HJ. Scoliosis after thoracoplasty. *J Bone Joint Surg (Am)* 1966;**48**:339–48.

22 Loynes RD. Scoliosis after thoracoplasty. *J Bone Joint Surg (Am)* 1972;**54**:484–98.

23 Shelton JE, Julian R, Walburgh E, Schneider E. Functional scoliosis as a long term complication of surgical ligation for patent ductus ateriosus in premature infants. *J Pediatr Surg* 1986;**21**:855–7.

24 Bisgard JD. Thoracogenic scoliosis. Influence of thoracic disease and thoracic operations on the spine. *Arch Surg* 1934;**29**:417–45.

25 Shands AR, Eisberg HB. The incidence of scoliosis in the State of Delaware. *J Bone Joint Surg (Am)* 1955;**37**:1243–9.

26 Winter RB. Spinal problems in pediatric orthopaedics. In: Lovell WW, Winter RB, eds. *Pediatric orthopaedics.* Vol 2. Philadelphia: JP Lippincott, 1986:606.