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Unusual Association of Congenital Atresias of the Oesophagus and Tricuspid Valve: Report of a Case

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Summary

Folami, A.O., Johnson, A., Smith, J. A., and Itayemi, S.O. (1978). *Nigerian Journal of Paediatrics*, **5** (**2**), 33. **Unusual Association of Congenital Atresias of the Oesophagus and Tricuspid Valve: Report of a case.** A case of congenital oesophageal atresia and distal tracheo-oesophageal fistula associated with tricuspid atresia is reported. To our knowledge, this rare association is the sixth case to be reported in the world literature and the first from the African continent.

Oesophageal atresia with or without tracheo-oesophageal fistula is frequently associated with other congenital malformations, the commonest being cardiovascular anomalies (Waterston, Carter, and Aberdeen, 1962; Holder *et al.*, 1964; Mellins and Blumenthal, 1964; Mehrizi, Folger and Rowe, 1966; David and O'Callaghan, 1974; Greenwood and Rosenthal, 1976). Though many different types of congenital cardiovascular malformations have been described co-existing with oesophageal atresia and tracheo-oesophageal fistula anomalies (Haight, 1957; Waterston, Carter and Aberdeen, 1962; Mellins and Blumenthal, 1964; Mehrizi, Folger and Rowe, 1966; David and O'Callaghan, 1974; Greenwood and Rosenthal, 1976), the association of tricuspid atresia with oesophageal atresia and tracheo-oesophageal fistula is very rare and only five cases have been reported previously (Haight, 1957; Mehrizi, Folger and Rowe, 1966; Silver *et al.*, 1972; David and O'Callaghan, 1974; Greenwood and Rosenthal, 1976). This communication documents the sixth such association.

Case Report

Baby A., UCH No. 388878, a male, was the product of a full-term pregnancy complicated by polyhydramnios and premature rupture of membranes. Delivery was uneventful. He was referred on the fifth day of life to the department of Paediatrics, University College Hospital, Ibadan with the provisional diagnoses of tracheo-oesophageal fistula and right upper lobe pneumonia.

On admission, he weighed 2.8 kg., was very active and acyanosed. Excessive mucus secretions in the mouth were observed. A nasogastric tube passed through the nose did not go beyond 9 cm. from the nasal orifice. Breath sounds were diminished in the right upper zone anteriorly. In the cardiovascular system the peripheral pulses were palpable, regular and of good volume. Heart rate was 140 per min. There was no clinical evidence of cardiac enlargement. Cardiac ausculation revealed a grade 3/6 pansystolic murmur, maximal in the left lower sternal border. X-rays of the neck and chest

showed the nasogastric tube stopping in upper oesophagus, right upper lobe opacity, normal pulmonary vascularity and a normal cardiac outline. Electrocardiogram (ECG) showed sinus rhythm, a rate of 120/min and QRS axis of +60. Abdominal X-rays revealed normal air distribution in the gastrointestinal tract. On the basis of the above clinical, ECG and radiological features the following problems were indentified:

1. oesophageal atresia
2. tracheo-oesophageal fistula
3. ventricular septal defect and
4. right upper lobe pneumonia.

He was treated with antibiotics, intravenous fluids and blood transfusion in preparation for surgery which was carried out two days after admission.

At surgery, oesophageal atresia with distal tracheo-oesophageal fistula was confirmed (Fig. 1a). The fistula was divided and closed and an end-to-end anastomosis was performed. Twenty-four hours after the operation he was feeding satisfactorily through a naso-gastric tube left in-situ at surgery, but unfortunately the baby pulled out the tube on the fourth post-operative day. A feeding gastrostomy was therefore performed. Subsequent clinical course was unsatisfactory with the development of empyema and a breakdown of the oesophageal anastomosis. He died on the 16th post-operative day.

Autopsy Findings

The body was that of a male neonate weighing 2.3 kg. and measuring 49 cm. in length. He had a right-sided empyema, extensive purulent exudate in the posterior mediastinum and a defect in the posterior tracheal wall at its bifurcation. The surgical anastomosis of the oesophagus had completely broken down, the upper and lower oesophageal fragments having retracted leaving a 5 cm. gap.

The right atrium was not dilated. The foramen ovale was patent. There was no tricuspid valve or orifice. The right ventricle was very small and hypoplastic. The main pulmonary artery, its valve and main branches were normal.

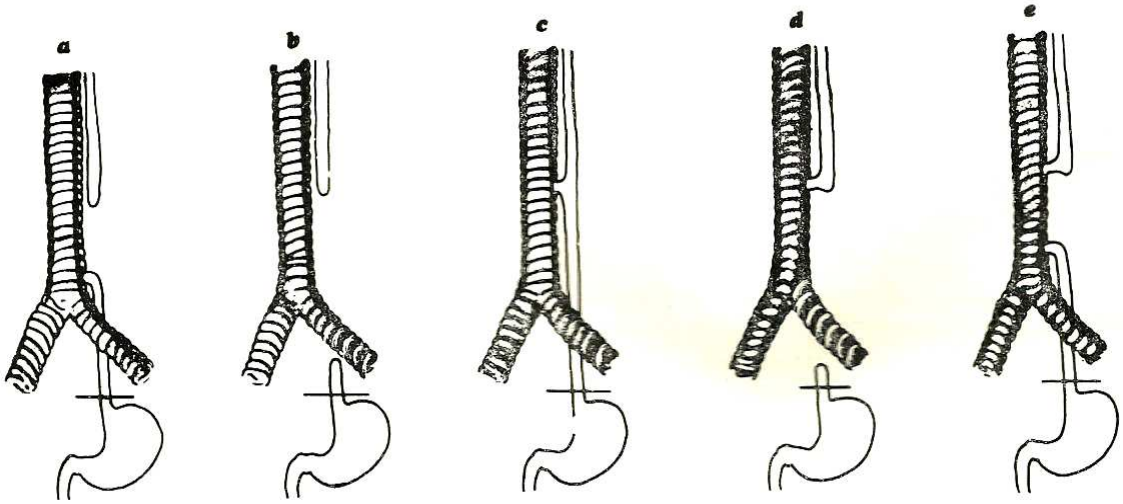

Fig. 1: Diagram showing five major anatomical types of congenital oesophageal atresia and tracheo-oesophageal fistula. Incidence of each type according to Holder et al., (1964) is 86.5 per cent for (a), 7.7 per cent for (b), 4.2 per cent for (c), 0.8 per cent for (d) and 0.7 per cent for (e).

The left ventricle was large and dilated. There was an oblique ventricular septal defect. The aorta and its valve were normal. There was no transposition of the great arteries, and the ductus arteriosus was not patent (Fig. 2). Histologically, the heart was unremarkable butlungs showed evidence of bronchopneumonia.

Discussion

The reported incidence of oesophageal atresia and tracheo-oesophageal fistula is about 0.3 per 1,000 births (Hamilton, 1969; David and O'Callaghan, 1974). Although several and confusing anatomical classifications of the malformations have been proposed (Keith, 1910; Vogt, 1929; Ladd, 1944; Swenson, 1948; Gross, 1953), there are however, five recognised types (Fig. 1) with few rare variants. By far the commonest type is oesophageal atresia with distal tracheo-oesophageal fistula.

The cause of oesophageal atresia and tracheo-oesophageal fistula is not clear. Developmentally, septation of the fore-gut into dorsal oesophagus and ventral trachea starts caudally in the fourth week of gestation. Tracheo-oesophageal fistula and indeed, oesophageal atesia may result from failure of the septation process although oesophageal atresia may in addition result from failure of complete re-canalisation of the oesophagus (Smith, 1957; Moore, 1973). The close proximity of the cardiac tube and fore-gut in early embryogenesis may explain why the commonest associated malformation is cardiovascular. Since anomalies of many other systems are associated also with oesophageal atresia and tracheo-oesophageal fistula, it is likely that various systems of the developing foetus are affected by a general insult. Aetiological factors in oesophageal atresia and tracheo-oesophageal fistula are heterogeneous and have been reviewed by David and O'Callaghan (1975).

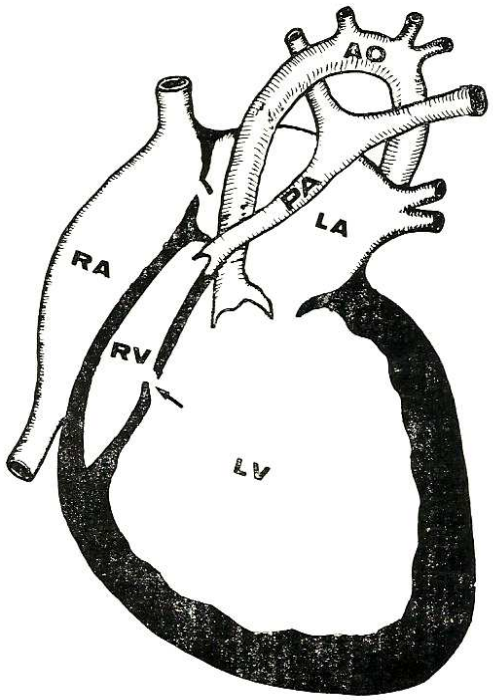

Fig. 2: Diagram illustrating the heart of the present case. Note the tricuspid atresia, hypoplastic right ventricle and an oblique VSD (arrow). RA = right atrium: RV = right ventricle; LV = left ventricle; LA = left atrium; PA = pulmonary artery; AO = aorta.

Other congenital malformations are frequently associated with oesophageal atresia and tracheo-oesophageal fistula, the reported incidences varying between 15 and 55 per cent (Waterston, Carter and Aberdeen, 1962; Holder *et al.*, 1964; David and O'Callaghan, 1974). In a large series reported by Holder *et al.* (1964), oesophageal atresia without tracheo-oesophageal fistula was the type most commonly associated with other congenital abnormalities (58 per cent), whilst tracheo-oesophageal fistula alone was the least (27 per cent). The percentage for oesophageal atresia with distal tracheo-oesophageal fistula, the type in our case was 48 per cent. The commonest associated malformations in oesophageal atresia and tracheo-oesophageal fistula are cardiovascular (Waterston, Carter and Aberdeen, 1962; Holder *et al.*, 1964; Greenwood and Rosenthal, 1976)

In the series reported by Greenwood and Rosenthal (1976) VSD was the commonest associated malformation when oesophageal atresia and tracheo-oesophageal fistula co-existed only with a cardiovascular anomaly, whilst Fallot's tetralogy was the commonest when there was additional gastrointestinal anomaly, and a complex cardiac anomaly was most commonly found when there was additional skeletal malformation.

Tricuspid atresia (TA) was the major associated cardiac malformation in the present case and this association has been reported previously in only five cases (Haight, 1957; Mehrizi, Folger and Rowe, 1966; Silver *et al.*, 1972; David and O'Callaghan, 1974; Greenwood and Rosenthal, 1976). TA is a serious and uncommon congenital malformation of the heart, the reported incidence in clinical and/or autopsy cases varying between 1 and 5.3 per cent (Wood, 1950; Campbell, 1955; Gasul *et al.*, 1960; Keith, Rowe and Vlad, 1967; Antia, 1974). The anatomical features of TA consist of four consistent malformations

(*i*) atresia of the tricuspid valve
(*ii*) interatrial communication
(*iii*) hypoplasia of the right ventricle and
(*iv*) hypertrophy of the left ventricle.

Early attempts at anatomical classification of TA included those of Kuhne (1906), Edwards and Burchell (1949).

A current and perhaps the most acceptable classification (Table I) is that proposed by Keith, Rowe and Vlad (1967). According to this classification, the present case coexisting with oesophageal atresia and distal tracheo-oesophageal fistula, belongs to type 1c (Fig. 2). In previous reports of TA coexisting with oesophageal atresia and tracheo-oesophageal fistula anatomical details were provided only in the case by Silver *et al.* (1972), which had TA (type-1b) and oesophageal atresia with distal tracheo-oesophageal fistula.

TABLE I

*Classification of Tricuspid Atresia and Relative Frequency of Anatomical Types in 143 Anatomical Specimens**

	Anatomical type	*Total Number of cases*	*Percentage of Total*
I.	Without transposition of the great arteries	99	69
	(*a*) Pulmonary atresia	13	9
	(*b*) Pulmonary hypoplasia Small ventricular septal defect.	73	51
	(*c*) No pulmonary hypoplasia. Large ventricular septal defect.	13	9
II.	With D-transposition of the great arteries.	40	28
	(*a*) Pulmonary atresia	3	2
	(*b*) Pulmonary or sub-pulmonary stenosis	11	8
	(*c*) Large pulmonary artery	26	18
III.	With L-transposition of the great arteries.	4	3
	(*a*) Pulmonary or sub-pulmonary stenosis	1	0.7
	(*b*) Sub-aortic stenosis	3	2

*Keith, Rowe and Vlad (1967)

In the present case the absence of cyanosis, a harsh grade 3/6 pansystolic murmur maximal in left lower sternal border, the absence of pulmonary oligemia led to the diagnosis of isolated VSD, a much commoner cardiovascular malformation than TA. Though difficulties like this sometimes arise in the diagnosis of TA, a careful review of electrocardiogram, echocardiogram (Kronzon *et al.*, 1975) and cardiac catheterisation will confidently make the diagnosis in life.

TA carries a poor prognosis, survival depending on adequacy of pulmonary blood flow which is related to the various anatomical types. According to Keith, Rowe and Vlad, (1967), 50 per cent of the cases are dead before the age of 6 months and 90 per cent by the age of ten years. A high mortality has been reported in oesophageal atresia and tracheo-oesophageal fistula associated with cardiovascular anomalies. David and O'Callaghan (1974) reviewing 345 infants with oesophaeal gatresia and tracheo-oesophageal

fistula showed that the mortality in those cases with cardiovascular malformations was 86.5 per cent; with no other malformation the mortality was 25.6 per cent and with additional non-cardiovascular anomaly it was 52.7 per cent. Greenwood and Rosenthal (1976) reviewing 326 infants with oesophageal atresia and tracheo-oesophageal fistula have also reported a mortality of 79 per cent among those associated with cardiovascular malformations and 25 per cent among those without. Palliative treatment of TA has been recently reviewed (Williams *et al.*, 1975; Williams *et al.*, 1976) and a radical physiological correction has been proposed (Fontan and Baudet, 1971), but it will be some years before the results of the latter procedure on survival may be properly evaluated.

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