Case Report

Non VACTERL oesophageal atresia, tracheoesophageal fistula associated with congenital hydrocephalus


Alzaiem Alazhari University*, Alribat University Hospital**, Al Hikma Private Hospital***, Khartoum, Sudan

Abstract
A case of uncommon association of non VACTERL oesophageal atresia, tracheoesophageal fistula and congenital hydrocephalus is described. It is hoped that it will raise the awareness to look for similar cases.

Keywords: VACTERL anomalies, oesophageal atresia, tracheoesophageal fistula, congenital hydrocephalus.

Introduction
Oesophageal atresia (OA) occurs in 1 in 3000 – 4500 livebirths. One third of patients are preterms(1-4). In 85% of cases there is a blind upper pouch and the lower pouch is joining the trachea just above the carina (type 1). At least 30% to 50% of cases have associated congenital anomalies cardiovascular in 25%, other digestive defects in 16%, urinary tract in 10%, skeletal in 10% and central nervous defect(1,2,3,4). About 10% of infants are associated with VATER or VACTERL anomalies (vertebral, vascular, anorectal, cardiac, tracheal, oesophageal, renal and limbs anomalies). VACTERL is associated occasionally with hydrocephalus and other defects(5,6,7,8).

This case report describes an uncommon association between oesophageal atresia (OA) and tracheoesophageal fistula (TOF) associated with congenital hydrocephalus.

Case report
A female baby was delivered at 35 week gestation by elective caesarean section due to congenital hydrocephalus which was diagnosed antenataly. This was the second pregnancy for a 29 years old mother. The outcome of first pregnancy was a normal child. Mother was attending regular antenatal care and ultrasound detected congenital hydrocephalus but no polyhydraminos. The baby cried at birth and birth weight was 2.5 kg. The head circumference was 45 cm corresponding to >97th centile with wide fontanel’s and suture diastases. In the ward, the baby was reluctant to breast feed and occasionally regurgitating. Attempts of passing a nasogastric tube failed and she was put on intravenous fluids.

Radiograph of the neck and chest plain and using dilute gastrographin contrast showed
an oesophageal pouch with the contrast stopping at the level of the second thoracic vertebra and with the presence of gas in the stomach (Fig. 1 and 2). Cranial ultrasound showed gross dilatation of all four ventricles with cerebral mantle of 15mm. The diagnosis of OA and TOF associated with congenital hydrocephalus was made.

The patient received the routine management of propping up and oesophageal suction. However, the patient developed signs of right sided aspiration pneumonitis which was confirmed clinically and radiologically (Figure 2) and due to it she succumbed on the third day.

**Discussion:**
To our knowledge this association of OA and TOF with congenital hydrocephalus was not described before in Sudan. The diagnosis of OA and TOF was confirmed radiologically (Figures 1 and 2). There was no clinical evidence of other VACTERL anomalies. It was reported that hydrocephalus is associated with VACTERL (5,6,7,8), but in this case it was only associated with OA and TOF.

The absence of polyhydraminos, the hydrocephalus with thin cerebral mantle, weak baby not showing excessive mouth secretions didn’t make the OA to be considered at birth. The possibility was raised after frequent vomiting and the failure to pass the nasogastric tube. Moreover, the attempts of feeding plus weak reflexes secondary to brain problem lead to the aspiration which caused pneumonitis. The signs of aspiration pneumonitis appeared clinically on the third day and was confirmed radiologically by the CXR that showed opacification of right hemithorax (Fig. 2) and this agrees to the literature(3).

The patient succumbed to aspiration pneumonitis before any surgical intervention. It was mentioned that at present in most developed countries the
presence of associated major congenital anomalies determines survival\(^9\). This however, is not the case in developing countries, where other factors continue to contribute to persisting high mortality\(^10\). Here both hydrocephalus with thin cerebral mantle and pneumonitis contributed to the death of the patient.

**Conclusion**

Although polyhydraminos in the mother may be the first sign of OA\(^4,8\). Yet many cases may not be associated with it\(^3\). Symptoms of OA might not be typical but immediate repeated vomiting after feeding may raise a suspicion. Hydrocephalus which may be associated with VACTERL\(^5,6,7,8\), can also be associated with only OA with or without TOF.

Patients in developing countries with such anomalies may die of other factors like lack of facilities, infections and aspiration pneumonia as well as by the presence of associated major congenital anomalies.

It is hoped that this patient being the first case described of such association in Sudan may raise the awareness to look for hydrocephalus associated with VACTERL or with only OA with or without TOF.

**References**


