

Vacterl association with hypertrophic pyloric stenosis

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How to cite this article:

Badebarin D, Aslanabadi S, Faramarzi M. Vacterl association with hypertrophic pyloric stenosis.

Iranian Journal of Pediatric Surgery 2018; 4(1):43 -46.

DOI: <http://dx.doi.org/10.22037/irjps.v3i3.13917>

Abstract

Keywords

- Vacterl
- hypertrophic pyloric stenosis
- pediatric

The acronym VACTERL is actually a combination of associated anomalies. We found a case of VACTERL association with hypertrophic pyloric stenosis. A six-weeks-old male infant was referred to us with hypertrophic pyloric stenosis. He had a history of esophageal atresia, imperforated anus and cardiac anomalies. This case shows hypertrophic pyloric stenosis in a patient with 'VACTERL' anomalies.

Introduction

VACTERL (Vertebral defects, Anorectal malformations, Cardiovascular defects, Trachea Esophageal defects, Renal anomalies, and Limb deformities) is a combination of associated anomalies that can be noticed on physical examination.^{1,2} If any one of these anomalies are found, we should look for other anomalies of this group. In this case report we introduce a case of VACTERL which also has hypertrophic pyloric stenosis (HPS).

Case Presentation

A male infant was referred to the neonatal intensive

care unit having respiratory distress beginning soon after birth. He had a blocked oral tube and imperforated anus at the first examination. He was born at 40 weeks of gestation by cesarean section to a 40-year-old mother. The prenatal history and teratogenic exposure was unremarkable and the birth weight was 3640 g. The parents were not related. Furthermore, the family history was unremarkable, particularly for malformations.

Our Physical examination revealed: drooling with excessive oral secretions, his oral tube was blocked 10-11 cm from the lips, lack of a midline gluteal fold and the absence of an anal dimple without any recto-perineal fistula. Further examination of other

received: 21 September 2016

accepted: 20 November 2016

ISSN 2423-5067

systems showed no more abnormalities.

We provided supplementary oxygen for the infant without artificial respiration. Alongside supplementary oxygen, we started antibiotic treatment.

Radiographic study revealed the presence of repleg tube in the upper pouch, GI air below the diaphragm and vertebral anomalies; hemivertebra in right side of L5 and left side of S2, which causes right lumbosacral kyphosis. On the echocardiographic examination, small closed PDA and mild TR

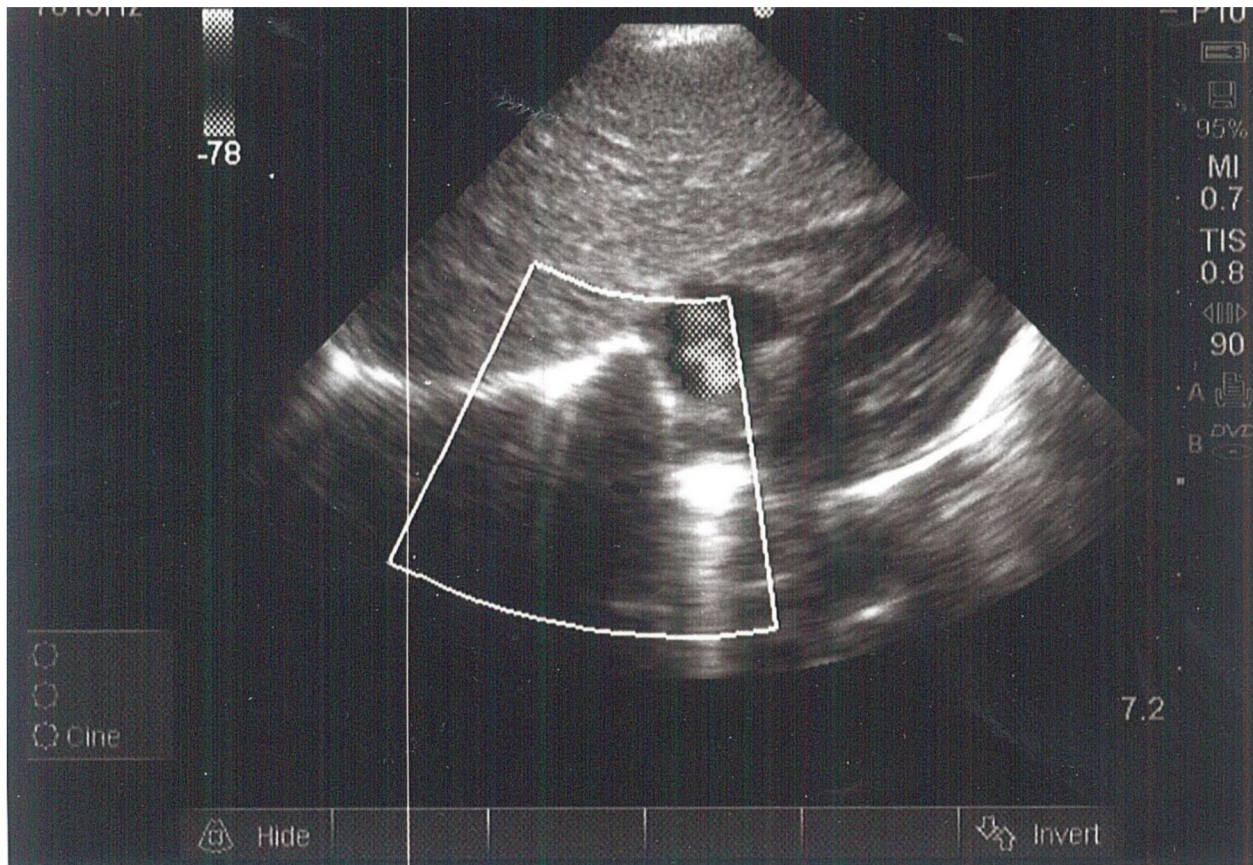


Figure 1: echocardiography demonstrating the small closed PDA and mild TR

and mild TR were present **Figure1**.

He underwent right postero-lateral thoracotomy and proximal to distal esophageal anastomosis. The tracheoesophageal fistula was cut and partial

colectomy with double barrel colostomy was performed.

When he was six-weeks-old, he developed failure to thrive and progressive vomiting and abdominal ultrasonography revealed signs of hypertrophic



Figure 2 : in operation room after pyloromyotomy, and his colostomy

pyloric stenosis (HPS). He underwent a successful pyloromyotomy **Figure 2**.

Discussion

Diagnosis of VACTERL is made when three or more of the six anomalies is seen.³ Our patient had four of the six anomalies involved in VACTERL, namely costovertebral (hemivertebra in right side of L5 and left side of S2), anorectal (imperforate anus), cardiovascular (mild TR), tracheoesophageal (esophageal atresia type C). Additionally, hypertrophic pyloric stenosis (HPS) was present.

In HPS muscular layers of the pylorus develops hypertrophy and hyperplasia which results in

functional gastric outlet obstruction. This pathology is the most common cause of gastric obstruction and surgical cause of vomiting in infants.

Although HPS following esophageal atresia repair is rarely reported, HPS with VACTERL complex has been reported only in one case with horseshoe lung and laryngeal cleft.⁴

The non-VACTERL anomalies seen in our case is probably another spectrum of the disease. Further reports are required to recognize non-VACTERL anomalies and clarify the etiologic relationship between them.

Conflict of Interest

There is no conflict of interest.

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