

CASE REPORT

It's not All Doom and Gloom: Prune Belly Syndrome Associated with VACTERL

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How to cite: Awad K, Lall A. It's not all doom and gloom: prune belly syndrome associated with VACTERL. J Neonat Surg. 2016; 5:33.

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ABSTRACT

Prune belly syndrome is a rare abnormality; its association with VACTERL is even rarer. This association has been reported in literature a few times since first reported in 1993 and so far the majority have either been stillbirths or died shortly after birth. We present a case of Prune belly syndrome associated with VACTERL who is now one year old.

Key words: Prune Belly Syndrome; VACTERL; Tracheo-esophageal fistula

INTRODUCTION

Serious congenital malformations cause early fetal loss as they are aborted or miscarried at an early gestational age. Two such examples of developmental defects are Prune belly syndrome (PBS) and VACTERL association. The concurrence of these two syndromes is extremely rare and is generally incompatible with life.[1-5] The incidence of 1.5% was reported for the occurrence of PBS with full VACTERL association and 17.5% for the three components of the VACTERL association [1]. No definite aetiology has been proven, but a defect in mesodermal differentiation, in early first trimester, has been suggested for these two syndromes [2]. We report a neonate of with VACTERL association who also had the features of prune belly syndrome.

CASE REPORT

A male neonate delivered by a normal vaginal delivery at 38 weeks of gestation presented for antenatally diagnosed hydronephrosis and maternal polyhydramnios. He required a very brief period of intermittent positive pressure ventilation and transferred to Special care baby unit after weaning off the ventilator. At birth, the following components composing of both VACTERL and Prune Belly syndrome were noted (Table1).

Table 1: Anomalies in Prune Belly Syndrome associated with VACTERL and their management

Anomaly	Management	Time of management
Tracheoesophageal fistula (TEF)	Right thoracotomy and Primary repair Esophageal dilatation	D1 of life 2 and 4 months
Anorectal malformation	Divided Colostomy	D1 of life
Bilateral impalpable Undescended testis	Planned	
Renal anomalies: Right renal agenesis , Duplex single left kidney with grade 4 vesico ureteric reflux (VUR) Abnormally dilated bladder with bilateral diverticulae Distal Hypospadias	Prophylactic Antibiotics CIC	D1 of life At 3 weeks
Musculoskeletal: Left club foot Right Tibial Agnesis Caudal Regression syndrome Sacral defects	Tenotomy	8 months
Respiratory: Tracheomalacia- severe	Aortopexy Tracheostomy	5 months
Right bronchomalacia	Non-invasive ventilation at night	6 months
Deficient abdominal wall muscles		
Gastrointestinal: Gastroesophageal reflux	Fundoplication and feeding gastrostomy	5 months

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Submitted: 16-02-2016
Conflict of interest: None

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Accepted: 20-04-2016
Source of Support: Nil

In summary, all the components of VACTERL association were present except cardiac defect. In addition, bilateral undescended testis; deficient abdominal wall muscles and urological anomalies formed the triad of prune belly syndrome.

He is now one year old, neuro-developmentally normal child, under the care of multidisciplinary team due to the complex nature of his anomalies. He is mainly gastrostomy fed, but takes small amount of pureed diet orally. He remains on 3-4 hourly CIC and prophylactic antibiotics. He is due for corrective hip surgery and bilateral orchidopexy this year.

Table 2: Previous case reports of Prune Belly Syndrome and VACTERL association

Year	Authors	Anomalies	Survival
1993	Reinberg et al (5)	VACTERL + Prune Belly syndrome	Stillbirth
1994	Ozturk et al (6)	VACTERL + Prune Belly syndrome	Unknown
1996	Lukusaet et al (3)	VACTERL + Incomplete Prune Belly syndrome in female patient	Unknown
2002	Potter et al (3)	VACTERL + Incomplete Prune Belly syndrome in female patient	Died
2004	Shah et al (4)	VACTERL + Incomplete Prune Belly syndrome in female patient	Died
2007	Ghritlaharey et al (2)	VACTERL + Prune Belly syndrome	Died D3
2008	Ely et al (7)	VACTERL + Pseudo Prune Belly Syndrome	Unknown
2012	Younes et al (3)	VACTERL + Prune Belly syndrome	Died D12
2014	Kar et al (1)	VACTERL + Prune Belly syndrome	Still birth

DISCUSSION

VACTERL association incidence is 1 in 10,000 to 1 in 40,000 [1], only 1.5% present with the full blown picture while 17.5% present with only 3 components of the association [2]. Prune belly syndrome is less common, with an incidence of 3.8 in 1,000,000 [1]. Hence the chance of having both these anomalies is extremely rare. There has been no solid evidence with regards to the etiology of

prune belly syndrome and some of the proposed theories are: Transient bladder outlet obstruction; the second theory is based on mesodermal arrest. Though transient bladder outlet obstruction may be able to explain findings of PBS, yet it fails to explain the VACTERL association. In our case we think the second theory could explain the possible association between PBS and VACTERL association [3].

This concordance of prune belly syndrome with VACTERL association has been reported a few times in literature as shown in Table 2. All the reported cases have been either still births or died shortly after birth. Our baby has survived his surgeries and is now one year old. He is neuro-developmentally normal and being followed up closely by a Multi-disciplinary team involving (Paediatric surgery and Urology, Orthopaedic surgery, Plastic surgery, Respiratory Physicians, Dietetics and Physiotherapists).

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