

Picture story

A case of omphalocele-exstrophy-imperforate anus-spinal defects (OEIS) complex

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Sri Lanka Journal of Child Health, 2019; **48**: 282-284

DOI: <http://dx.doi.org/10.4038/sljch.v47i3.8556>

(Key words: Omphalocele, bladder exstrophy, imperforate anus, spine, Sri Lanka)

Introduction

The OEIS complex comprises omphalocele, exstrophy of the cloaca, imperforate anus and spinal defects¹. It is due to incomplete closure of the ventral abdominal wall during early embryonic development². There are no published cases in Sri Lanka. We report a 3 month old infant with OEIS complex.

Case Report

A 3 month old girl was transferred from the paediatric surgical unit for further evaluation and management of multiple congenital abnormalities and recurrent ileostomy diarrhoea. She was the second child born to non-consanguineous parents following an uncomplicated antenatal period. The baby was born at 35 weeks of gestation with a birth weight of 1.95kg. At birth, she was found to have minor omphalocele, cloacal exstrophy and high lying imperforate anus. (Figure 1)

Antenatal ultrasound scans did not reveal any of these abnormalities. Rest of the clinical examination revealed a sacral lipoma, congenital talipes equinovarus deformity and overlapping toes in the right foot. (Figures 2 and 3)

There were no dysmorphic features in this child. X ray of the lumbosacral spine showed scoliosis and malformed underdeveloped caudal vertebrae (Figure 4).

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(Received on 04 October 2017: Accepted after revision on 24 November 2017)

The authors declare that there are no conflicts of interest

Personal funding was used for the project.

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Figure 1: 3 month old girl with OEIS complex



Figure 2: Sacral lipoma



Figure 3: Right sided clubfoot



Figure 4: Scoliosis and malformed caudal vertebra

Ultrasound scan of the spine revealed a tethered cord. 2D echocardiography revealed a small atrial septal defect, a tiny patent ductus arteriosus and mild pulmonary hypertension. She underwent first stage cloacal exstrophy repair with the creation of an ileostomy and bladder plate reconstruction surgery at the age of 2 days. Since six weeks of age, she presented with recurrent ileostomy diarrhoeal episodes requiring intravenous fluid and electrolyte therapy. She has severe failure to thrive and her current weight at the age of five months is 2.6kg.

Her haematological investigations revealed microcytic hypochromic anaemia with anisopoikilocytosis and low serum ferritin suggestive of iron deficiency anaemia. Her renal functions and ultrasound scan of kidney, ureter and bladder were normal. Diagnosis of OEIS complex was made on clinical grounds and follow up was arranged involving a multidisciplinary team.

Discussion

Although the exact etiology of OEIS complex is unknown there are reported cases in association with chromosomal disorders such as Edward syndrome, Down syndrome etc.⁴. However, in this case, dysmorphic features suggestive of a genetic syndrome were not present. OEIS complex has a wide spectrum of clinical abnormalities⁵. Although cardiovascular system involvement is unusual, a few cardiac abnormalities were found in this child. Though the diagnosis of the OEIS complex is evident at the time of birth, it is difficult to diagnose it antenatally⁶. Non visualization of the fetal bladder, defect in the infra-umbilical anterior abdominal wall, omphalocele and myelomeningocele are considered as the major criteria. The minor criteria include club foot, renal abnormalities, ascites, widened pubic arches, narrow

thorax, kyphoscoliosis, hydrocephalus and single umbilical artery⁷. Except for right sided club foot and scoliosis the other features were not present in this child.

Patients with the OEIS complex need a multidisciplinary approach and early surgical management is recommended in the neonatal period. Patients will have to undergo reconstructive surgery in several stages throughout life⁸. The prognosis of the OEIS complex is variable and is determined mainly by the severity of the structural defects. Whilst some deaths have occurred in infancy, some have survived till the second decade⁹.

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