International Journal of Sciences & Applied Research

www.ijsar.in

Embryonic decompression with vascular disruption - A case report

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Abstract

The present case report is regarding the incidental finding of the new born with numerous congenital malformation which includes: Hydrocephalous, Syndactyly, Cloacal Anomaly, Deformed upper and lower limb and dextrocardia .

The foetus was born to the mother through normal vaginal twin delivery; one baby was healthy and normal in weight and characters while the other baby had multiple congenital malformations.

Keywords: Dextrocardia, Syndactly, Decompression, disruption

Introduction

Embryo is one of the most dynamic biological systems and in contrast to adults, drug effects are often irreversible. The combination of embryonic compression with vascular disruption results in extensive extremity and body wall defects in humans have been termed "**LIMB BODY WALL COMPLEX**".

This defect occurs with a birth prevalence of 0.26 per 10000 births, with half of cases being still born, and have strong association with low birth weight, short gestational age nd younger maternal age. Lower limb more severely and consistently affected than upper limbs.

Case report

A rare case of multiple congenital malformation was born to a healthy mother with no significant complication during pregnancy. It was a twin delivery one baby was healthy and normal in weight and characters while the other baby had multiple congenital malformations which include Hydrocephalous, Syndactyly, Cloacal Anomaly, Deformed upper and lower limb and dextrocardia (2,3,4,5).

The new born showed hydrocephalus which is due to inability of an infant to adequately control C.S.F. volume resulting into increase in ventricular size with subsequent rise in intracranial pressure leading to potential neurological complication and subsequently death. The neonate also showed low set ears and hypertelorism.

Syndactyly: It is fusion of digit or toe varies from a cutaneous web to a synostosis and may occur. As an isolated trait or as a component of certain syndromes like apert syndrome, carpenter syndrome, fetal hydantoin syndrome, orofascial digit syndrome.

Cloaca is undifferentiated and there is a single perineal opening.

There is deformity of upper extremity with bilateral shortening and bilateral syndactyly.

The lower limb is also shortened with sealed limbs.

X-Ray Findings: The radiograph shows the shadow of liver on left side, splenic shadow and stomach shadow on right side showing situs inversus(8,9,10).

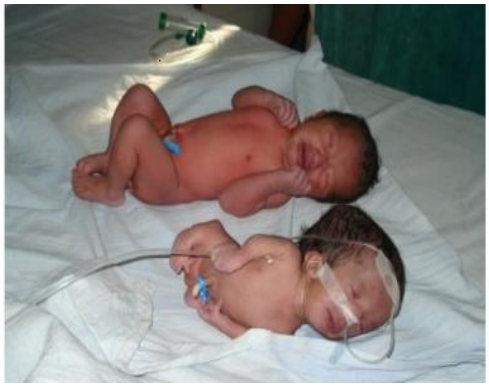

Figure 1

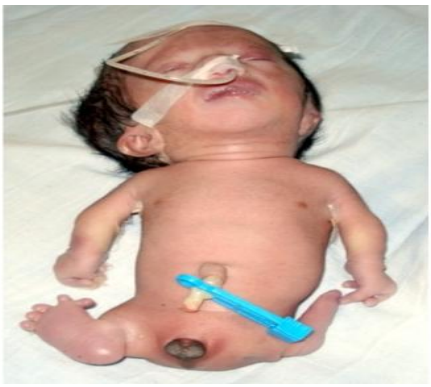

Figure 2

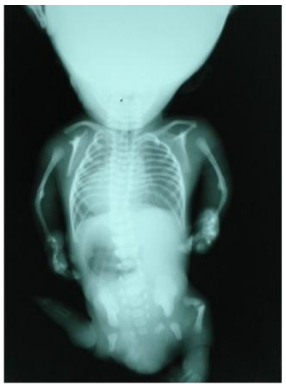

Figure 3

It also shows dysmorphic spine and single forearm bone with scoliotic spine, hemi-vertebrae T11 – T12 with absent 11^{th} and 12^{th} rib on left side. The femur and tibia are absent on right side with absence of ankle and foot bone while on left side there is shortening of femur with single leg bone. tarsals are not well defined while metatarsals on left side are well defined.

Discussion

Constraint that occurs during the latter period of gestation cause moulded deformations with good prospects for spontaneous or assisted return to normal form. The type of defects produced by such compression during early period of organogenesis (within first trimester) fall under three category

1. moulded deformations
2. incomplete morphogenesis
3. dysruption of morphogenesis

The combination of early embryonic compression with vascular disruption results in extensive extremity and body wall defects in humans has been termed limb body wall complex. This defect occurs with a birth prevalence of 0.26 per 10000 births with half of cases still born and have strong association with very low birth weight, short gestational age and younger maternal age. The spectrum of defects includes variable combination of body wall defects with evisceration of thoracic and abdominal organs, limb deficiencies, neural tube defects, and facial clefts with or without amniotic bands and renal agenesis. Lower limb more severly and consistenly affected than upper limb with distal structures more involved than proximal. Infants with limb reduction defect due to early exposure to misoprostol or chorionic villus sampling have asymmetric digit loss , constriction rings and syndactyly as a result of vascular disruption in limb structures that had formed normally.

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