

## Picture Story

# Congenital varicella syndrome with jejunal atresia

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*Sri Lanka Journal of Child Health*, 2010; **39**: 154-155

(Key words: Congenital varicella syndrome, jejunal atresia)

## Introduction

Varicella zoster virus (VZV) infection in pregnancy leads to infection of the fetus since VZV is capable of crossing the placental barrier at any time during gestation<sup>1,2</sup>. Maternal varicella in the first trimester is responsible for multiple malformations of offspring with characteristic clinical features involving brain, eye, skin, musculoskeletal and gastrointestinal systems<sup>3,4</sup>. We present a neonate with congenital varicella syndrome and jejunal atresia.

## Case report

A baby girl was born to a 19 year old unmarried mother who had varicella infection around 12 weeks of gestation. An emergency caesarean section was done for fetal distress at a gestational period of 36 weeks and 4 days. The birth weight was 1.465 kg, Apgar score 6 at 1 minute and 10 at 5 minutes.

She was admitted to the premature baby unit due to poor general condition & the presence of large haemorrhagic necrotic denuded skin lesions of both lower limbs. Antenatal scan at 21 weeks had not revealed any fetal anomalies.

The multiple skin lesions comprised healed scars with both hyperpigmented and hypopigmented areas along with new haemorrhagic and necrotic skin lesions. The leg below the right knee joint was almost devoid of skin and muscle with circumferential scarring (cicatrix). Right gastrocnemius muscle was hypoplastic and right foot had a talipes equinovarus deformity (Figure 1).

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There was paucity of movements of both lower limbs. Bilateral femoral pulses were present with the distal limb being viable. X ray of right lower limb showed tibial constriction. She also had right side microphthalmia (Figure 2).

Ophthalmological referral showed cataract on right side and corneal scarring on left side. However, the ultrasound scan (USS) of the brain was normal.

On the first day of life the apathetic hypotonic neonate developed bilious vomiting with abdominal distension following expressed breast milk feeds given via nasogastric tube (Figure 3).

Urgent USS of the abdomen revealed upper gastrointestinal (GI) obstruction, gaseous distension with minimal amount of fluid in bowel loops, ascites and hyper-echoic areas in the liver with possible aerobilia (Figure 3).

Urgent exploratory laparotomy found two small jejunal perforations, one jejunal loop being dilated with distal obstruction. Dilated loop was excised and end to end jejunal anastomosis done. Baby was ventilated for two days postoperatively and extubated on day 4 of life after the successful outcome of the surgery.

2D echocardiogram showed fenestrated inter-atrial septum and a small atrial septal defect for which indomethacin therapy was suggested but not given since platelet count was  $28 \times 10^9/L$ .

Plastic surgical referral and reconstructive surgery as well as cataract surgery was planned in one month's time.

(Received on 20 September 2010. Accepted on 29 October 2010)



**Figure 1:** Skin lesions & talipes equinovarus



**Figure 2:** Microphthalmia



**Figure 3:** Abdominal distension

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