Picture Story

Prune belly syndrome

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Sri Lanka Journal of Child Health, 2008: **37**: 30

(Key words: Prune belly syndrome, Eagle-Barrett syndrome, Triad syndrome)

A term, 4 kg baby boy was born by elective caesarean section. Antenatal ultrasound had shown oligohydramnios, lung hypoplasia and ascites. There was an antenatal history of gestational diabetes mellitus and pregnancy induced hypertension.

He had gasping respiration and bradycardia at birth, an Apgar score of 2 at one minute and no chest expansion on bag ventilaton. He was intubated and ventilated but did not show any improvement and died at 30 minutes of age.

Abnormalities noted at birth included a large distended lax abdomen with deficient musculature, distended bladder, facial flattening with hypoplasia of right side, hypoplastic right chest with air entry more on the left side, right undescended testis and bilateral club feet (Figure 1).

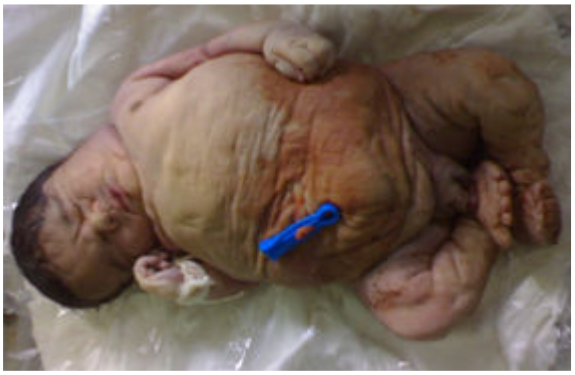

Figure 1 *Appearance of baby at birth*

Post mortem examination revealed hypoplastic kidneys, left megaureter, megacystis, right sided intra-abdominal testis (Figure 2) and bilateral pulmonary hypoplasia (Figure 3).

Baby was diagnosed to have Prune belly syndrome also known as Eagle-Barrett syndrome and Triad syndrome[1]. It has an incidence of 1 in 40,000 and 95% affected are male[1]. The characteristic association of deficient abdominal muscles, undescended testes and urinary tract abnormalities probably results from severe urethral obstruction in fetal life[1]. Oligohydramnios and pulmonary hypoplasia are common perinatal complications[1].

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(Received on 20 December 2007. Accepted on 27 December 2007.)

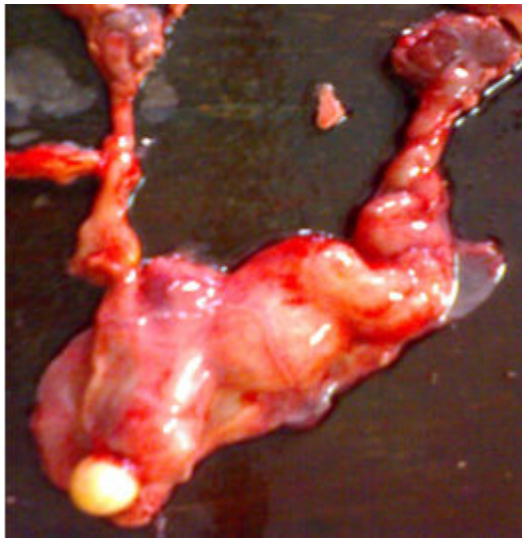

Figure 2 *Bilateral hypoplastic kidneys, left mega ureter, megacystis and an intra-abdominal testis*

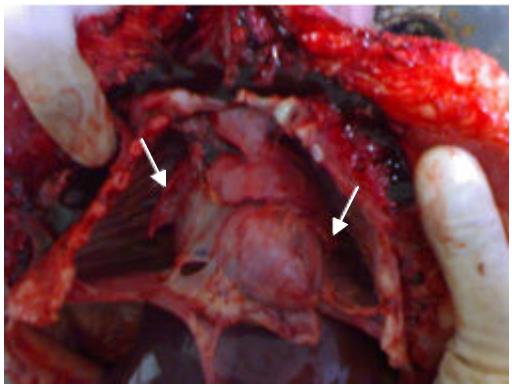

Figure 3: *Bilateral pulmonary hypoplasia*

Reference

1. Kliegman RM, Behrman RE, Jenson HB, Stanton BF. editors, Nelson Textbook of Paediatrics. 18th ed. Philadelphia: Saunders, 2007; 2240.