Prune belly syndrome

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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).

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 syndrome¹. It has an incidence of 1 in 40,000 and 95% affected are male³. The characteristic association of deficient abdominal muscles, undescended testes and urinary tract abnormalities probably results from severe urethral obstruction in fetal life¹. Oligohydramnios and pulmonary hypoplasia are common perinatal complications¹.

Reference