A case of Potter syndrome due to posterior urethral valve

In February 2004, a baby boy (birthweight, 3.6 kg) was delivered at full term at the Kwong Wah Hospital by vacuum extraction because of prolonged second stage of labour. No antenatal care had been sought by the parents—a non-consanguineous couple who had already had a healthy child. The baby exhibited dysmorphic features, such as hypertelorism, prominent epicanthic folds, a flat and broad nose, low-set ears, a receding chin, and flexion contracture of the limbs. Endotracheal intubation and mechanical ventilation were administered because breath sounds were diminished for both lungs, the baby grunted and had difficulty crying, and he also had cyanosis. The chest X-ray (Fig) showed bilateral pneumothorax and lung hypoplasia, and also revealed a distended bladder, which suggested bladder outlet obstruction. Renal ultrasonography showed bilateral hydronephrosis and gross hydroureters. After chest intubation and administration of surfactant, ventilation remained difficult and oxygen saturation could not be maintained despite a high inspiratory pressure (up to 55 cm H_2O) and high-frequency oscillatory ventilation. The echocardiogram, however, was normal and showed no evidence of persistent pulmonary hypertension. A Foley catheter was inserted to drain the distended urinary bladder. The urine sodium level was high (116 mmol/L) and the serum creatinine concentration was markedly elevated (115 μmol/L)—indications of impaired renal function. The baby died of respiratory failure after 17 hours of life. Investigations for septics gave negative results, and chromosome analysis revealed a normal 46,XY karyotype. Post-mortem examination showed posterior urethral valves, bilateral hydroureters, bilateral hydronephrosis, obstructive renal dysplasia and bilateral lung hypoplasia—features compatible with the diagnosis of Potter syndrome.

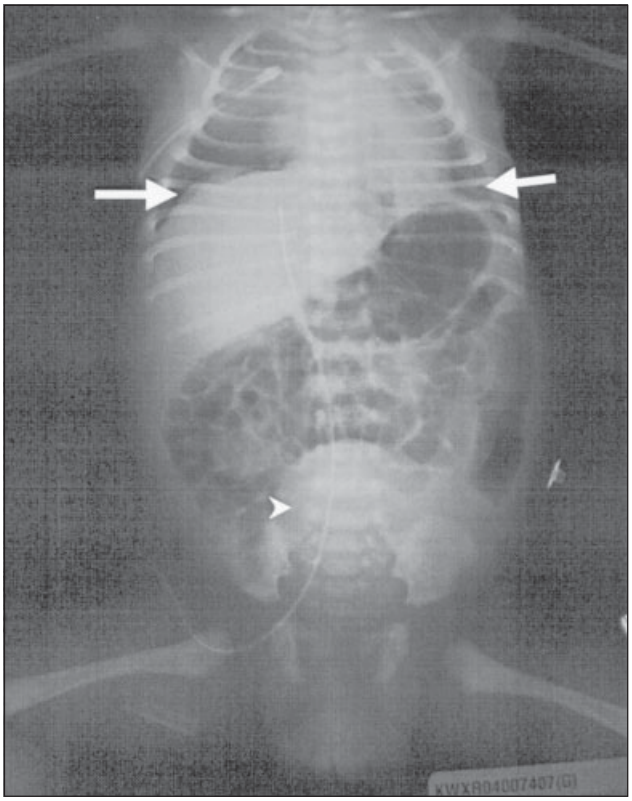

Fig. X-ray showing bilateral lung hypoplasia and pneumothorax (arrows), chest tubes in situ, and a grossly distended urinary bladder (arrow head)

In a review of 80 cases of Potter syndrome, common abnormal renal findings were bilateral renal agenesis (21%), cystic dysplasia (47%), and obstructive uropathy (25%).[1] The presence of posterior urethral valves is one of the most common causes of bladder outlet obstruction, accounting for more than 60% of cases in a 6-year series in Glasgow.[2] During embryogenesis, the most caudal end of the mesonephric duct is absorbed into the primitive cloaca at the site of the future verumontanum in the posterior urethra. In healthy males, the remnants of this process are the posterior urethral folds (plicae colliculi). Fusion of these primitive folds which have an abnormally high position of insertion is believed to be the origin of 95% of the posterior urethral valves. Obstruction of urine flow through the urethra into the amniotic fluid gives rise to severe oligohydramnios, dilatation of the bladder, ureters and even the kidneys, as in our case. The mechanism by which oligohydramnios produces lung hypoplasia is not completely understood. It is widely accepted that for lung development to proceed normally, physical space in the fetal thorax must be adequate and amniotic fluid must be brought into the lung with fetal breathing movements. Thus, it follows that extrinsic compression of the fetal thorax causes hypoplasia, either by inhibiting respiratory movements or by forcing out fluid from the lungs. The latter hypothesis was supported by Nicolini et al,[3] on the basis of an observed reduction in amniotic pressure. Hence, the instillation of artificial amniotic fluid could restore abnormal amniotic pressure and help prevent pulmonary hypoplasia. The combination of amnio-infusion and bladder aspirations has achieved a successful outcome in the management of obstructive foetal hydronephrosis.[4]

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References

1. Curry CJ, Jensen K, Holland J, Miller L, Hall BD. The Potter sequence: a clinical analysis of 80 cases. Am J Med Genet 1984;19:679-702.
2. Rani R, Cameron A, Munro F. Prenatal diagnosis and management of urethral obstruction. J Obstet Gynaecol Res 1997;23:59-62.
3. Nicolini U, Fisk NM, Rodeck CH, Talbert DG, Wigglesworth JS. Low amniotic pressure in oligohydramnios—is this the cause of pulmonary hypoplasia? Am J Obstet Gynecol 1989;161:1098-101.
4. Tabsh KM, Theroux NL. Obstructive fetal hydronephrosis managed by amnioinfusions and bladder aspirations. Obstet Gynecol 1997; 90:679-80.