Anaesthesia for prune belly syndrome

Adrian Bösenberg, *MBChB, DA(SA) FFA (SA)*
Department of Anaesthesia, University of Cape Town, South Africa

Synopsis of patient: An 18month old, 12.8kg male presented for laparotomy. He is a known 'prune belly' syndrome and had undescended testes. The purpose of the laparotomy was to locate his testes and perform a bilateral orchidopexy if possible. His mother gave a history that he suffered from recurrent chest infections but his chest was clear on the day of surgery. He had no cardiac abnormalities. His abdominal musculature was deficient and had no tone. It seemed to consist of folds of loose redundant skin (Fig1a and b). On straining or coughing the abdominal wall was very protuberant. (Fig 2). He was known to have bilateral hydronephrosis with compromised renal function. His electrolytes were within normal limits and the urea slightly elevated. Haemoglobin was 9.8gm. (Haematocrit 28). His clotting profile was normal. No premedication was given. He received halothane anaesthesia supplemented with an epidural at T11-12. Relaxants were not considered necessary. An epidural infusion of 0.2% bupivacaine at 1.5ml/hour was used for 48 hours and the postoperative course was uneventful.

PRUNE BELLY SYNDROME

Prune belly syndrome is rare with an estimated occurrence of 1:40000 births. The name is derived from the characteristic wrinkled appearance of the abdomen that resembles a prune. The syndrome usually consists of a triad of congenital anomalies - deficiency of the abdominal wall musculature, cryptorchidism and urinary tract anomalies.[1-3] The syndrome is not confined to this triad and may also involve the respiratory, cardiovascular, skeletal and gastrointestinal systems in 75% of affected individuals.[1-3]

More than 95% are boys and there is no obvious racial predilection. Females usually only have abdominal wall involvement. The prognosis may vary from death in utero or a normal life expectancy.[2] Survival depends on the severity of the renal and pulmonary impairment with 20% dying within the first month of life and 50% within 2 years. The syndrome may also be referred to as Eagle-Barrett syndrome, Triad syndrome or abdominal musculature syndrome.[1,2]

Although prune belly syndrome is considered a distinct entity by most investigators, there is no consensus as to its pathogenesis despite extensive study of clinical cases and pathological material.[2] The embryological theory suggests that

Correspondence:
Professor A Bosenberg
email: bosie@cormack.uct.co.za

Figure 1a and 1b: Typical wrinkled skin of the infant's abdomen at rest under anaesthesia.

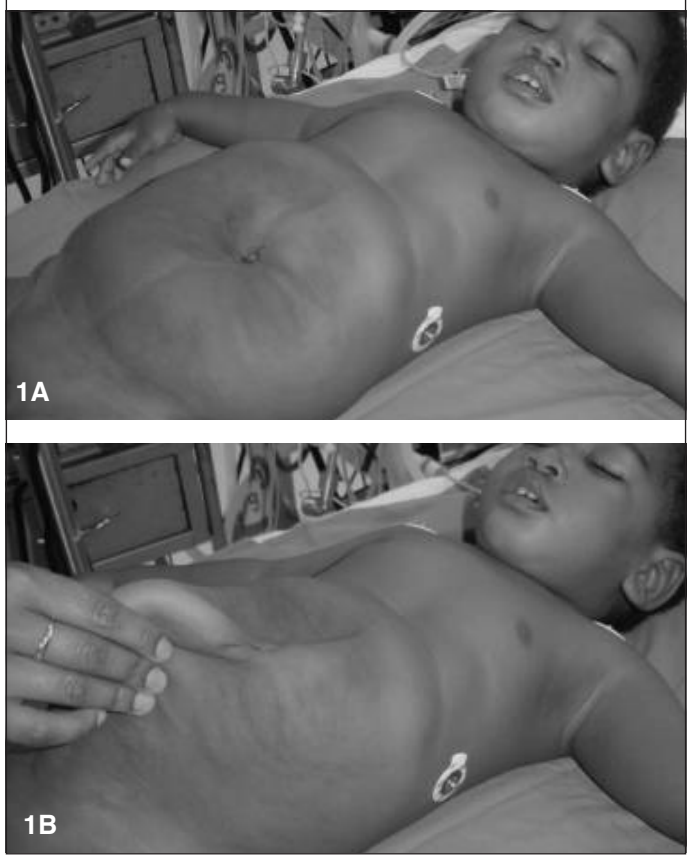
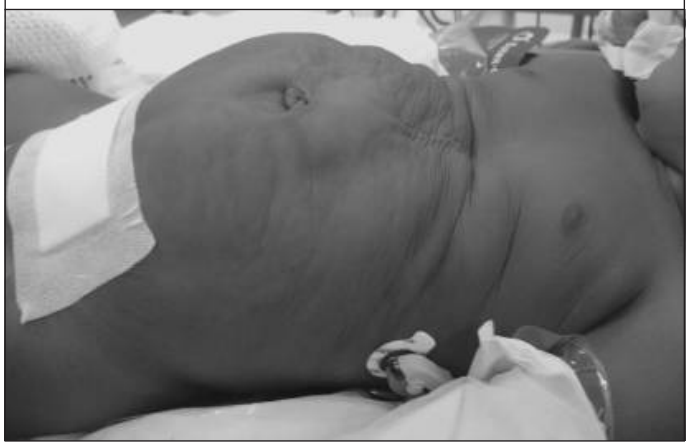


Figure 2: Abdominal appearance of the same infant while coughing prior to extubation.



there is an aberration of mesenchymal development at around six weeks gestation whilst others consider the distal urinary tract obstruction as the primary underlying mechanism. A vascular aetiology is a third proposal, which attempts to explain the lower limb abnormalities, but these are probably secondary to iliac vessel compression by the dilated urinary tract rather than a primary vascular insufficiency.

Cardio-respiratory manifestations: Pulmonary hypoplasia may occur as a consequence of oligohydramnios caused by the distal urinary tract obstruction in utero. If severe, the pulmonary hypoplasia may lead to stillbirth or demise in the early neonatal period.

In those presenting for surgery respiratory complications are the leading cause of postoperative morbidity.[3] These children have an ineffective cough because of the deficient abdominal musculature and the relative flat diaphragm. Pectus excavatum may compound the problem.[4] The risk of respiratory infection is increased as a result. Opiate analgesics should therefore be used with caution, and regional anaesthesia is considered the best option.[4]

Laryngeal atresia[5] and difficult intubation[6] have been described in association with this syndrome. The airway should be secured rapidly wherever possible, to reduce the risk of regurgitation.

Cardiac defects may be seen in approximately 10% of affected individuals. The most commonly encountered defects are similar to those seen in children with cardiac defects in the general population i.e. patent ductus arteriosus, atrial septal defects, ventricular septal defects and Tetralogy of Fallot. Prophylactic antibiotic cover is advised.

Orthopaedic manifestations: The prevalence of musculoskeletal problems requiring orthopaedic intervention is in the order of 45%.[7] The involvement may be congenital or developmental. Congenital anomalies affect the lower limbs in particular and include club feet, limb deficiencies, hip dysplasias and vertebral malformations. The suggested mechanism for these is compression of the iliac arteries by the obstructed urinary tract. The developmental defects may be secondary to renal disease (renal osteodystrophy) or include scoliosis and pectus carinatum or pectus excavatum.[4] These defects support a mesenchymal defect.

Genito-urinary manifestations: Distal urinary tract obstruction is usually not detectable clinically, suggesting a functional rather than a physical cause for the obstruction. This may lead to severe dilatation of the bladder and ureters causing bilateral hydronephrosis. Kidneys are often small and dysplastic. Affected individuals are prone to urinary tract infections, increasing the likelihood of renal failure. Preoperative evaluation of renal function is therefore important. Bilateral undescended testes are characteristic. The urinary tract anomalies, if present, are not as severe in affected females.

Gastro-intestinal manifestations: Chronic constipation may be a presenting feature. Malrotation and imperforate anus are also common presenting problems. Reflux may present an additional anaesthetic risk.

Anaesthetic considerations: In view of the abnormal abdominal musculature, ventilatory support should be mandatory as spontaneous ventilation may be significantly impaired by general anaesthesia. Muscle relaxants are probably not necessary in view of the lax abdominal wall. The response to muscle relaxants is said to be normal but compromised renal function may affect elimination. All drugs dependant on the kidneys for excretion should be used with caution.

Some authors advise the use of neuromuscular blockers when ventilating these patients to reduce the risk of pneumothorax or pneumomediastinum particularly in those individual with pulmonary hypoplasia.[3] All respiratory depressants should be used with caution.[3,8]

Conclusion

Although this syndrome is rare, a knowledge of the pathophysiology involved is essential to plan an appropriate anaesthetic.[8] Perioperative management can go a long way to preventing serious morbidity or even death.[3]

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