General Anesthesia for a Case of Abdominal Musculature Deficiency, Prune Belly, Syndrome

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The prune belly syndrome (PBS) is characterized by the absence of musculature in the lower central and medial part of the abdominal wall, cryptorchidism and urinary tract anomalies[1-3]. Anesthesia is thought to be hazardous in the PBS due to the deficiency of abdominal muscles, which could possibly lead to respiratory difficulty both intra- and postoperatively. To our knowledge, however, there are only a few reports on anesthesia in the PBS[4-6]. This is a case report of a child with the PBS, discussing the problems which can arise during the anesthetic management of this condition.

Case Report

A boy aged 6 years, 19 kg in weight and 111 cm in height, was admitted to our hospital for the right nephrectomy. The patient had already been suspected to have the PBS in the fetal period upon an ultrasonic examination. The child was born at 35 weeks of gestation, weighing 3,000g, by Cesarean section. Since he was born, the abdomen had been noted to be lax and wrinkled, and the bilateral undescended testicles were observed. These features were indicative strongly of PBS.

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The child had undergone the repeated surgical procedures, i.e., the cystostomy at the day of his birth, the right ureterostomy at the 42nd day of his life, and the release of ileus at age 4 showing no malrotation of the intestine. All of them were performed under general anesthesia without any significant complication. This time, the right nephrectomy was indicated not only for the recurrent pyelonephritis but also for the deterioration in quality of life, because the incontinent urine fluxed continuously through the ureterostoma.

The general condition of the child was found to be quite good at the pre-operative visit. The belly was protuberant in the upright posture (figs. 1 and 2) and fell backwards in the supine position. The costal margin was everted and the subcostal angle was wide. Both testicles were undescended. The thoracic cage was deformed as pectus carinatum.

Except in very quiet respiration there was obvious activity in the accessory respiratory muscles. In any particular respiratory effort, for instance during crying, there was slight indrawing of the lower ribs on inspiration. The child was not in respiratory distress and no cyanosis was noted. The respiratory rate at rest was 20/min. The chest X-ray was almost normal but showing the diaphragm to be slightly flat, which is common in the PBS. The pulmonary function test was not performed.

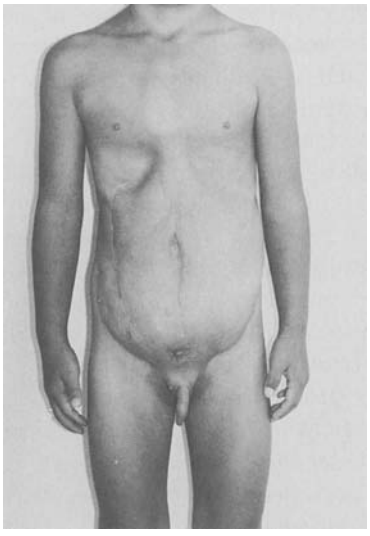

Fig. 1. Frontal standing view, which shows the protuberant abdomen, the eversion of the costal margin and the widening of the subcostal angle. The undescended testicles, the cystostoma, the right ureterostoma through which the urine is fluxing, the deformity of thoracic cage and the scar of operation for the ileus are also shown.

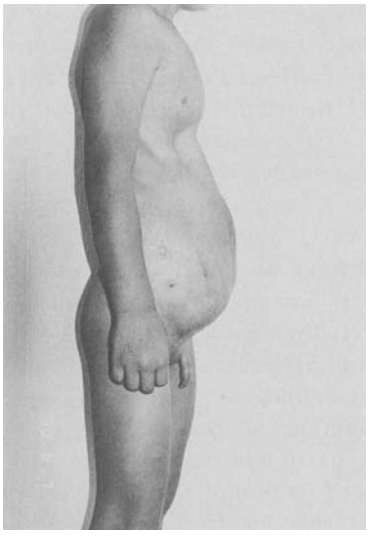

Fig. 2. Lateral standing view.

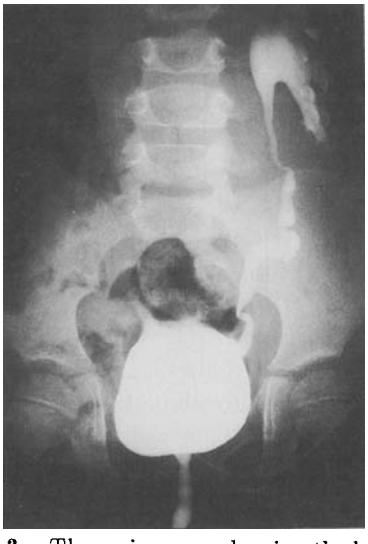

Fig. 3. The vesicogram showing the left vesicoureteric reflux during micturition.

An echogram showed the right hydronephrosis. In an intravenous pyelogram, the right renal pelvis could not be visualized. The vesicography revealed the left vesicoureteric reflux, Grade II a; low pressure type, during micturition (fig. 3). The creatinine clearances measured with the urine obtained from the bladder and the right ureterostoma were 44.2 and 14.0 $l \cdot day^{-1}$, respectively. Other laboratory examinations showed the normal values.

The child was satisfactorily premedicated with subcutaneous atropine (0.2 mg) and hydroxyzine (20 mg) given 45 min prior to the induction of anesthesia. Following peripheral intravenous cannulation, anesthesia was induced with thiamylal (150 mg) and orotracheal intubation was performed with an endotracheal tube (5.5 mm ID without a cuff) using suxamethonium (20 mg). Anesthesia was maintained with nitrous oxide/oxygen 3:2 $l \cdot min^{-1}$ and halothane 1.5–1.0%. Ventilation was manually controlled during operation. Throughout the procedure, there were no abnormalities in the respiratory sound on the anterior chest wall and in Sa_{O_2} measured with pulse oxymetry. The blood pressure and heart rate were stable. The rectal temperature was kept normal (36.6–37.3°C) by using a heating pad. No muscular relaxants were given except suxamethonium for endotracheal intubation. The surgical procedure was performed in the left lateral position and lasted 2 hr 35 min. The total blood loss was about 40 ml. A 460 ml of Solita T1™ was given over 3 hr 55 min of the total

anesthesia time. The child made an uneventful recovery from anesthesia. The arterial blood gas values were pH 7.34, Pa_{O_2} 470 torr, Pa_{CO_2} 41 torr and base deficit 4.0 mEq·l^{-1}, 10 min after the extubation in spontaneously breathing oxygen (5 $l \cdot min^{-1}$) by a face mask.

Oxygen was given as a routine precaution in the early postoperative period. Particular attention was paid to respiratory physiotherapy during the subsequent period to avoid retention of secretions and the development of atelectasis. No significant postoperative complications were observed, though the excretion of bowel gas delayed until the 3rd postoperative day. The patient was discharged in good condition on the 20th postoperative day. In the near future, urological procedures are scheduled to correct the vesicoureteric reflux and the undescended testes.

Discussion

The congenital deformity characterized by a wrinkled abdomen was originally described by Fröhlich in 1839[2,7]. The term of prune belly was aptly coined for the syndrome by Osler in 1901[7]. Agenesis of abdominal muscles, gross urinary tract malformations and undescended testicles are the main features of the PBS. The abdominal wall deformity may vary in degree from a decrease to complete absence of abdominal wall musculature[7,8]. In some cases there are further congenital abnormalities, which include malformation of the heart, malrotation of the gut, deformity of the thoracic cage, talipes equinovarus, scoliosis, skeletal deformities and gross mental deficiency.

The syndrome is limited almost entirely to males. The incidence may be 1/35,000–50,000 births[9]. Though little is known about the etiology, one possible theory relates to a chromosomal abnormality[7,8]. As a result of the severity of the disease and its complications, the mortality rate may be as high as 50% before the age of two years depending on the number of physical abnormalities present[9].

The PBS is uncommon, but individual cases may frequently require repeated surgical procedures. The absence of abdominal muscles weakens the ability to cough vigorously, and consequently there is a tendency to recurrent respiratory infections. Moreover, the problems of the abnormal mechanics of pulmonary ventilation in this condition may confront anesthesiologists.

Some reports described the difficulties in respiratory management of PBS, i.e., preoperative recurrence of infection, paradoxical respiration during anesthesia, and postoperative respiratory failure which required support by positive pressure ventilation[4,5]. Henderson et al. reviewed 36 children with PBS who underwent 133 operations[6]. In their cases, postoperative respiratory tract infections followed eight of the 133 anesthetics and three deaths occurred in the postoperative period. One death was related to the sequelae of the PBS, i.e., the severe aspiration bronchopneumonia secondary to the repeated vomiting, possibly associated with uremia.

If there is much impairment of renal function, particular care has to be taken in the administration of drugs and intravenous infusions. In the presence of uremia, respiration may be further complicated by the development of a special type of pulmonary edema, sometimes called uremic pneumonitis.

Caution should be exercised to avoid respiratory embarrassment in the use of premedication and postoperative analgesics known to produce respiratory depression. The weakness of the abdominal muscles may partly cause the delay of the postoperative excretion of bowel gas as seen in our case. The PBS may show hip dysplasia and dislocations[7,9]. Proper intraoperative positioning of the patient must be advocated, particularly in the lithotomy position, where hip dislocations are likely to occur.

In summary, a 6-year-old boy suffering from the PBS was anesthetized for the nephrectomy without any significant complication. The problems of anesthesia in association with the PBS are discussed.

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