Pathophysiologic and anesthetic correlations of the prune-belly syndrome

JOYCE P. HOLDER, CRNA, MA
Brooklyn, New York

"Prune-Belly" is the name given to the disease which is characterized by a congenital wrinkled appearance of the abdomen. Usually, a triad of congenital anomalies highlights the components of the prune-belly syndrome. This triad consists of undescended testicles, abdominal musculature deficiency and urinary tract abnormalities.

The previously described triad of the syndrome is by no means the total spectrum of the disease. Prune-belly syndrome is also associated with diseases of the respiratory, cardiovascular, skeletal, gastrointestinal and central nervous systems. Associated diseases of major body systems which are caused by renal and cardiopulmonary anomalies, as well as the surgical procedure to be performed, influence the plan for the administration of anesthesia. The nature of the genitourinary diseases in the prune-belly baby may obviate the need for surgical intervention as early as the neonatal period of life.

Although this congenital disease occurs with some rarity, the gravity of the syndrome demands an in-depth knowledge of its pathophysiology in order to assure uncomplicated anesthetic care. Astute surveillance during preanesthetic, anesthetic, and postanesthetic management is germane to the prevention of mishaps.

The congenital deformity characterized by a wrinkled and deformed abdomen was originally described by Frohlich in 1839. In 1895, Parker noted a collection of three anomalies: the undescended testicles, urological obstructive diseases and abdominal wall deficiency. The term "prune-belly" was aptly coined for the syndrome by Osler.

Within succeeding years and following numerous urological studies, a myriad of nomenclature evolved. Other names for the syndrome include the triad syndrome, abdominal musculature deficiency syndrome, Eagle-Barrett syndrome, mesenchymal dysplasia, and congenital absence of the abdominal musculature syndrome. (See Figure 1.)

Incidence and etiology

Reports have indicated that the prune-belly syndrome occurs in 1-35,000-50,000 births. Males are more frequently affected than females, with the females accounting for about 5% of the cases reported. 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.

The etiology of prune-belly syndrome has not yet been decided upon, however, there are several existing theories for its occurrence. One such theory relates to a chromosomal abnormality. Opinions describing embryological defects have also been theorized by Nunn and Stephens. Subsec-
system obstruction, and renal dysplasia. Radiographic evaluation has proven to be a very important tool for providing a description of the prune-belly syndrome. Megaloureteres, ureteral obstruction, dilated bladder and hydronephrosis are some of the associated diagnoses which are discovered through radiographic studies.

It must not be misconstrued that the triad of congenital deformities related above presents the total description of the prune-belly syndrome. In many instances a diagnosis of prune-belly syndrome may be accompanied by a few or many other abnormalities including skeletal, respiratory, gastrointestinal, cardiovascular, and central nervous system defects.

One of the skeletal deformities which may be found with prune-belly syndrome is Potter's facies in which there is a receding chin, malpositioned ears and a flattened nose. Bilateral club feet, as well as dislocations and dysplasia of the hips, arthrogryposis, and accessory digits may be present. Torsion of the spine and pectus excavatum may also be noted.

Abnormalities of the gastrointestinal tract in the prune-belly baby are related to malrotation of the intestines. Other gastrointestinal conditions found to be concomitant with the syndrome are Hirschsprung's disease, imperforate anus and omphalocele.

Descriptive pathophysiological elements

There are three commonly occurring components of the prune-belly syndrome. The prune-belly appearance resulting from abdominal wall deficiency, undescended testes, and urinary tract malformation are the major anomalies of this congenital disease (Figure 2).

The abdominal wall deformity may vary in degree from a deficiency to a complete absence of abdominal wall musculature. When there is very little abdominal musculature to support the contents of the abdomen, layering of the intestines can be seen beneath the skin. In extreme cases, peristalsis may also be observed.

Urological disorders of the prune-belly syndrome range from simple to complex urinary and renal abnormalities. Throughout the genitourinary tract, congenital malformations may be found, for example, dilatation of the urinary tract, urinary tract imperfection, malformation of the yolk sac and an initial obstruction of the urinary system, have also been listed.

Abnormalities of the gastrointestinal tract in the prune-belly baby are related to malrotation of the intestines. Other gastrointestinal conditions found to be concomitant with the syndrome are Hirschsprung's disease, imperforate anus and omphalocele.
Evaluation of the cardiovascular system may show malformation of the heart.9,11 About 10% of individuals may be diagnosed as having cardiac anomalies along with prune-belly syndrome.12 Fallot's tetralogy and septal defects are the cardiac conditions usually found.4,5 Patent ductus arteriosus occurs with less frequency.4

Welch and Kearney reported findings of abnormalities of the central nervous system; specifically cerebral hypoplasia and premature ossification of the cranial sutures.4

Congenital pulmonary hypoplasia also has been related to the prune-belly syndrome.1,3,6 The weak or absent abdominal muscles, chest deformity, flaring outward of the lower ribs1,6,8 and lack of accessory muscles of respiration8 are all features which have negative effects on pulmonary ventilation. The prune-belly infant has little or no coughing ability because of abdominal muscle weakness.1,2,8,11 This disability has resulted in pneumonia, recurrent respiratory infections and atelectasis. Congenital cystic adenomatoid malformation of the lung was described by Weber as a concomitant condition with prune-belly syndrome.13

Preanesthetic implications

Management of the prune-belly patient requires meticulous anesthetic considerations in order to avoid complications. Patients with this syndrome undergo a myriad of surgical procedures. These procedures may be necessary either to diagnose or to treat certain situations, and in many instances require anesthesia intervention. Frequently, the prune-belly baby initially needs genitourinary surgery. At one end of the spectrum, anesthesia may be needed for simple procedures such as meatotomy; at the other end, ileal loop, cystectomy

The abdominal muscles, ribs and diaphragm play an important role in the mechanics of pulmonary ventilation.14 (See Figure 3.) Therefore, a deficiency in the abdominal muscles will inhibit pulmonary ventilation, because the normal contraction of muscles is reduced and movement of the diaphragm is limited. Indeed, a preanesthetic physical assessment of the respiratory system is necessary to discover whether there is a need for preoperative respiratory toilet or pharmacological treatment. A history of the incidence of cyanosis will indicate problems

![Figure 3](image-url)

with oxygenation or severe cardiopulmonary compromise.

A preanesthetic assessment of the laboratory data is mandatory. Emphasis is placed on serum creatinine (normal 0.6-1.3 mg/100 ml) and creatinine clearance (normal 85-140 ml/min) values, which are substantial indicators of renal status. Serum creatinine may increase as much as tenfold in renal disease.\(^\text{14}\) Advancement of urological problems in the prune-belly patient may lead to severe renal disease, which may also be evidenced by increased serum potassium levels (normal 3.5-5 mEq/L). Knowledge of this information gleaned during physical assessment will assist in the preparation of an appropriate plan for anesthetic management.

### Anesthetic correlations

Induction of anesthesia for the prune-belly baby may be a tedious task considering the vast number of anomalies which are associated with the prune-belly syndrome and the major body systems which could require surgical intervention within the early weeks of life. The anesthetic technique chosen will depend on the pathophysiology of the individual patient plus the surgical procedure to be performed. A selection of various types and sizes of face masks is necessary, since the possible presence of Potter's facies may prevent a snug mask fit. Adequate oxygenation is germane to an uneventful anesthetic induction for the prune-belly patient.

Inhalation induction is the most commonly used technique for pediatric patients when a vein cannot be easily cannulated. Halothane may be chosen over enflurane and isoflurane as an inhalation agent. During induction, enflurane and isoflurane may cause breath holding or laryngospasm because of their more pungent odor. Hannington-Kiff pointed out that the prune-belly infant should be treated as a vomiting risk patient since respiration is embarrassed and the ability to cough is weak.\(^\text{11}\) Thus, the prune-belly patient is very likely to aspirate quietly during induction and emergence.

The better method of airway management is by tracheal intubation, as the airway may need intermittent suctioning to clear secretions. In addition, associated conditions such as pectus excavatum or atelectasis which restrict respiratory volumes may require positive pressure ventilation. Ideally, an awake intubation is recommended if the muscles are extremely flaccid. Whenever possible, muscle relaxants should be avoided. Given the associated cardiac and respiratory problems which are described with the prune-belly syndrome, it is advised that as many monitoring devices as possible be instituted before induction. Pulse oximetry, for example, is an excellent indicator of oxygen saturation and should be applied from preinduction through recovery periods while managing the prune-belly baby.

Vigilant monitoring of the cardiopulmonary status is of paramount importance to early detection and treatment of problems occurring during anesthesia. Carbon dioxide monitoring and blood gas analysis would provide indices to show the effectiveness of ventilation. During anesthesia maintenance, muscle relaxants should be used sparingly, if at all, and their effects monitored with a nerve stimulator. The prolongation of a neuromuscular blockade, which could be caused by the interaction of muscle relaxants and aminoglycoside antibiotics,\(^\text{15}\) is noteworthy since the prune-belly patient may be receiving this type of antibiotic therapy for recurrent infections of the genitourinary tract.

As is customary, continuous temperature monitoring is mandatory. Hypothermia must be avoided in the pediatric patient. A warm room, overhead radiant heat, thermal blanket and wrapping the infant in a plastic drape are methods of maintaining body temperature. Ryan et al. recommended that room temperatures should be 80°F (26.6°C) for newborns and small neonates; 78°F (25.5°C) for infants up to six months of age; and 76°F (24.4°C) for patients six months to two years of age.\(^\text{17}\) Irrigation fluids, when used by the surgical team, must be warmed to prevent lowering of the patient's temperature.

Fluid therapy should follow the prescribed standards and guidelines for pediatric patients. Nevertheless, for the prune-belly baby it must be remembered that if renal failure is present, there must be a reduction in the volume of fluids administered.

Proper positioning of the prune-belly patient must be advocated, especially in the lithotomy position, where hip dysplasia and dislocations are likely to occur because of the previously discussed skeletal anomalies.

Postanesthesia management of the prune-belly infant requires prudent judgment and skill. Whenever possible, the infant should be transferred to the recovery room with the endotracheal tube in place. Not only is the prune-belly patient prone to develop postanesthesia respiratory insufficiency\(^\text{10}\) and may require mechanical ventilation, but chest physiotherapy and intermittent tracheal suctioning\(^\text{1, 2, 11}\) will assist in the prevention of pulmonary complications.
Before the patient is extubated and prior to discharge from the recovery room, a chest x-ray and blood gas analysis should be used as measures of detecting possible atelectasis and respiratory insufficiency. The extubated prune-belly infant should be placed in a position which will allow drainage of airway secretions and freedom of chest movements. An oxygen tent or humidified oxygen can be invaluable in facilitating ventilation. On arrival in the recovery room, the application of an apnea monitor is sagacious action for calling attention to respiratory difficulties.

Summary

Although the prune-belly congenital anomaly occurs with rarity, the associated pathophysiology is so severe that appropriate anesthesia management requires in-depth knowledge of this complex syndrome. Each patient requires individual evaluation, as the degree of anomalies varies from patient to patient. Indeed, an application of the anesthetic correlations to affected body systems of the prune-belly infant will limit possible complications.

REFERENCES


AUTHOR

Joyce P. Holder, CRNA, MA, is a graduate of Harlem Hospital School of Anesthesia for Nurses, New York. She received a BS degree in Health Care and Administration from St. Francis College, New York, and an MA in Community Health from Brooklyn College in New York. Ms. Holder is currently a clinical assistant professor for the State University of New York, College of Health Related Professions in Brooklyn. She also is a staff nurse anesthetist at State University of New York Health Science Center at Brooklyn, and director of Kings County Hospital Center, School of Anesthesia for Nurses.

ACKNOWLEDGMENT

The author wishes to thank Ms. Ellen L. Jackson for manuscript preparation.