

The Orthopaedic Manifestations of Prune-Belly (Eagle-Barrett) Syndrome*

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ABSTRACT: Forty children were managed for prune-belly syndrome between 1979 and 1989, and twenty-five of them had musculoskeletal abnormalities. The musculoskeletal abnormalities were primary in twenty-two children, secondary to renal osteodystrophy in one, and both primary and secondary to renal osteodystrophy in two. Thirteen children had marked abnormality of the hip, and congenital dislocation was typically resistant to conventional treatment. Scoliosis was seen in seven patients. Pectus excavatum (a chest-wall deformity) was seen in eleven patients, including five of the six who had an idiopathic-like curve. Although prune-belly syndrome is uncommon, the diagnosis necessitates a thorough orthopaedic evaluation because of the high prevalence of associated musculoskeletal abnormalities.

Prune-belly syndrome is characterized by a triad of abnormalities, consisting of cryptorchidism, obstructive uropathy, and absent or hypoplastic abdominal muscles¹¹. This group of clinical findings was apparently first described by Osler in 1901. The condition was later defined as a distinct clinical syndrome by Eagle and Barrett in 1950. The syndrome occurs primarily in boys^{1,2,4,12-15}, and the prevalence has been estimated as one in every 50,000 live births⁵.

The name prune-belly syndrome is derived from the characteristic wrinkled prune-like appearance of the belly of the neonate, which is the result of the profound deficiency of abdominal muscles (Fig. 1). The genitourinary involvement includes a wide range of abnormalities of the kidneys, ureters, and bladder^{1,2,6,10,12,13,15}. Cryptorchidism is a consistent finding in male patients. Mortality is generally related to the severity of the pulmonary and genito-urinary abnormalities.

While the musculoskeletal abnormalities have been described previously in the pediatric and urological literature^{1,2,4,6,12}, to the best of our knowledge the orthopaedic

literature contains only two reports addressing this syndrome^{7,14}. The purpose of the current study was to characterize and to determine the prevalences of the musculoskeletal abnormalities in our patients who had prune-belly syndrome.

Materials and Methods

The medical records and radiographs were reviewed retrospectively for a consecutive series of forty patients who had been managed for prune-belly syndrome at The Children's Memorial Hospital in Chicago between 1979 and 1989. There were thirty-eight boys and two girls. The average age at the time of presentation at our institution was two years and four months (range, birth to fifteen years) (Table I), and the average age at the most recent follow-up examination was seven years and four months (range, birth to nineteen years and nine months). Thirteen of the forty patients had been followed at our institution since birth. The patients had been hospitalized an average of six times (range, one to twenty-eight times) for various genito-urinary, respiratory, and orthopaedic problems related to the prune-belly syndrome. Four patients (Cases 8, 9, 30, and 32) died of respiratory failure with postmortem documentation of pulmonary hypoplasia, in the early months of life. There were no other deaths in the current series.

The average gestational age was thirty-seven weeks (range, thirty to forty-one weeks). Fifteen of the forty patients were neonates (one month old or less) at the time of presentation at our center. Fourteen of twenty-five patients for whom the history of delivery was known had been delivered by cesarean section. The average birth weight of the forty patients was 3.0 kilograms (range, 1.7 to 3.9 kilograms), which was below the twenty-fifth percentile. Thirty-two newborns were below the twenty-fifth percentile for height and thirty were below the twenty-fifth percentile for the circumference of the head. No patient had a sibling who had prune-belly syndrome.

Results

Musculoskeletal Abnormalities

The prevalence of musculoskeletal abnormalities in all forty patients was 63 per cent (twenty-five of forty)

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