Management of the Abdominal Wall Defect in the Prune Belly Syndrome by Muscle Transportation

An 18-Year Follow-up
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Prune belly syndrome (PBS), an uncommon anomaly, consists of genito-urinary abnormalities and a partial or complete absence of abdominal wall musculature. Although the patient's genito-urinary problems are addressed, the attention currently directed toward the abdominal wall deficiency has been mostly aimed at improving the cosmetic appearance of patients and does little to replace the important functions of the musculature. These functions are the support of the viscera and their compression as well as the movement of the trunk. A case report with an 18-year follow-up is presented where thigh muscles were transposed to act as substitutes for the missing musculature. Although this presentation is based on a single case, it is intended to alert patients and physicians to a method of improving the lot of patients with PBS. In this patient, the result was very satisfactory. From being unable to move his trunk as a schoolboy before the surgical procedure, postoperatively he was able to participate in all school activities. As an adult, he is fully active and is a working musician. His excretory functions are normal and the scoliosis, present when first seen, has not progressed.

It is suggested that consideration be given to the management of the abdominal wall deficiency in PBS by the transportation of muscle(s), because no alternative effective treatment directed to restoring absent muscle function appears to exist.

Because the operative procedure is well tested and successful in adult patients who lose their abdominal wall musculature, the dissemination of the data appears to be necessary. Clin. Anat. 13:341-346, 2000. 2000 Wiley-Liss Inc.

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hypoplasia antero-lateral abdominal wall muscles; reinforcement by thigh muscles; long-term follow-up

INTRODUCTION

Prune Belly Syndrome (PBS) is an uncommon syndrome almost exclusive to males and is characterized by a variable aplasia of the abdominal wall musculature associated with undescended testes and urinary tract anomalies of varying degrees. The theories proposed as to the cause of malformation are many and mostly involve an early mesodermal defect whose center appears to lie in the first lumber segment, from which the greater part of the transverse and oblique muscles develop with the hypoplasia diminishing cranially and expressing themselves as occasional lower limb defects and a frequent absence of the upper part of the rectus abdominis (Gray and Skandalakis, 1972). Whereas the prognosis for life is related to the severity of the uropathology, the quality of life is closely related to the degree of developmental failure of the abdominal musculature.

Treatment in the past was almost exclusively directed toward the urinary tract pathology (Nunn and Stephens, 1961; Gray and Skandalakis, 1972; Raffensperger, 1990; O'Neill et al., 1998), but more recently attention has turned toward improving the appearance of, and to a considerably
lesser degree the function of, the abdominal wall musculature (Duckert, 1976; Randolph et al., 1981; Erlich et al., 1986; Montfort, 1991).

A patient with all the components of the PBS and who underwent surgery for the abdominal wall defect is presented.

CASE REPORT

A 13-year old male presented with a history of having undergone numerous urologic procedures for genito-urinary abnormalities. He had been hospitalized seven times for problems related to lack of abdominal musculature. His main complaint at this time was difficulty in moving his trunk. As a result, his activities became so limited that he shunned attending school; as he put it, he could not sit up without help. He had been wearing a brace for some years in attempt to control the scoliosis that had ensued, and backache was a continual problem.

Physical examination revealed a boy who, apart from being somewhat overweight, was of normal height and development. Abdominal examination showed a protuberant abdomen that bulged into the loins (Fig. 1). The superfluous skin was floppy and healed scars in the midline and costo-vertebral areas were present. Using specifically designed exercises (Kendall and McCreary, 1983), it was not possible to appreciate any active abdominal wall musculature. Testes were present in the scrotum. Clinical and radiological evidence of a marked rotational thoracolumbar scoliosis were demonstrated.

The boy and his parents were aware that muscle transposition procedures were being carried out for the management of recurrent incisional and inguinal hernias (Ger and Duboys, 1983; Ger et al., 1984). They requested that this approach be used in this case. The patient was instructed to carry out a course of exercises to strengthen and develop the muscles of the antero-lateral thigh muscles before the proposed surgery.

OPERATIVE TECHNIQUE

A midline lower abdominal incision that excised the old scar was made. The absence of musculature as far as the midaxiillary lines was confirmed. Vertical and oblique incisions in the right thigh and a vertical incision on the left thigh were fashioned. After incising the deep fascia, the rectus femoris muscles were dissected out and the tendons divided just superior to the patellae. The sartorius muscles were then mobilized and the tendons of insertion divided, the right side requiring an additional oblique incision to gain adequate access. On the right side, the descending branch of the lateral circumflex artery was carried out as far as proximally at the point of entry of its nerve supply. The four muscles were then passed deep to a skin bridge anterior to the inguinal ligaments (Fig. 2). As a small deficiency still existed in the upper abdomen, the rectus femoris muscles were lengthened by multiple incisions in their posterior sheaths, a maneuver that allowed complete closure of the defect. The muscles, placed in the following sequence from left to right; left rectus femoris, left sartorius, right sartorius and right rectus femoris, were sutured to each other and anchored by #0 Dexon sutures. The skin of the abdomen, now excessively superfluous, was partly excised and the wound drained. The postoperative course was uneventful.
Following discharge, the patient continued physio-therapy to his lower extremities and was seen intermittently by his pediatric surgeon (E.C.). His appearance had improved considerably (Fig.3), and in a short while he was able to sit up by himself and take part in physical activities at school. Contact with the patient was resumed 18 years later (R.G.). Now 31 years of age, he reported that he was doing well at work and was also a working musician. Physical examination at this time revealed a young man who had no relevant complaints and was in good general health. The abdominal wall showed a reasonable profile and a good ability to contract; a transposed muscle was visible and palpable (Fig. 4). He was able to maintain the semireclined position by means of his abdominal wall muscles (Fig. 5). Extension of the knee joints was full and active against resistance. An x-ray examination of the spine showed that there had been no progression of the scoliosis, the radiographic study being a mirror image of the study 18 years earlier.

DISCUSSION

In earlier management of PBS, emphasis fell mainly on renal problems, and the abdominal deficiency was dealt with conservatively by support in the form of elastic bandages and corsets (Gray and Skandalakis, 1972), especially when it was observed that some patients improved with the passage of time (Nunn and Stephens, 1961; Raffensperger, 1990; O'Neill et al., 1998). Gradually operative procedures were introduced commencing with plications (Duckett, 1976), followed by excisions of the lower 5-7.5 cm of the abdominal wall in cases where it appeared that the musculature of the lateral and upper parts of the abdominal wall were present to some degree as judged by electromyography (Randolph et al., 1981). More extensive procedures were then attenuated and hypoplastic musculo-fascial layers were dissected out and overlapped in a double-breasted manner, which resulted in considerable cosmetic and psycho-social improvement. This procedure was modified by Montfort et al. (1991) who used the full thickness of the abdominal wall that was considered to produce a better cosmetic result. References were made by Ehrlich et al. (1986) and Parrott and Woodard (1992) to an operative approach described by Ger (1983) whereby certain muscles of the antero-lateral thigh were transposed to the abdominal wall; they also noted that long-term results were unavailable. In the latter context it can be pointed out that the muscle transposition procedures, introduced in 1966 (Ger), have been part of the ongoing management of various lesions in a considerable number of sites in the body. These areas include the abdominal wall, and the lesions vary from incisional hernias (Ger and Duboys, 1983), recurrent inguinal hernias (Ger et al., 1984) to recalcitrant infectious and destructive conditions (Ger, in press). The muscles used to treat lesions of the abdominal wall included the rectus femoris, sartorius, gracilis, and tensor fascia lata. These muscles were chosen because they had free borders that did not require sharp separation from adjacent muscles, such as occurs when one of the other components of the quadriceps femoris such as the vastus lateralis was occasionally used. The number of muscles used depends on the site and size of the muscular defect. In the case presented, the defect was long and wide and required four muscles.

The transportation of skeletal muscles differs conceptually from the other operative procedures described above whose main aim is to improve the cosmetic and psycho-social situation. The muscle transposition approach attempts to supplant the missing actions of the antero-lateral abdominal wall muscles. These muscles support, and, by rapid contraction, protect the abdominal viscera from injury. With the aid of a closed glottis, muscle contraction raises the intra-
abdominal pressure sufficiently to empty abdominal contents such as feces, urine, gastric contents, and a baby at childbirth. The muscles initiate forward bending of the trunk, assist rotation of the spine, and prevent the latter's hyperextension. It follows that significant effects may accompany the failure or underdevelopment of the abdominal muscles. As the latter play a major role in spinal movements, especially those of rotation, it is clear that muscular deficiencies or imbalances, especially of the two oblique muscles, are likely to result in the development of scoliosis. There do not appear to be any references in the literature concerning the prevention or limitation of this serious problem. Although it is difficult to substantiate that the muscle transposition procedure played a major part in the cessation of this patient's scoliosis, it remains a distinct possibility because "structural curves that occur in myopathic cases have a strong tendency to progress during the adolescent growth spurt" (Turek, 1984).

Synthetic prostheses, popular in the management of acquired defects of the abdominal wall, do not appear to have been used in this congenital condition. Their general disadvantages are well known, but in this situation, additional unfavorable features are the obvious lack of contractility and the inability to keep pace with the growing patient. The advantages of muscles as the agents of repair are that they are dynamic structures that are effective in controlling abdominal wall herniae (Ger and Duboys, 1983), they may relieve the symptoms and even present or limit scoliosis, improve backache (Toroanto, 1988), and assist bowel and urinary actions by potentiating expulsive abdominal efforts. In this operative approach, there should also be no difficulty in retaining the umbilicus, a cosmetic consideration stressed by Montfort (1991).

This presentation falls into the category of a case report since the current system of medical practice in the United States makes it extremely unlikely that a case of PBS would be referred to a general surgeon; the patient would be cared for by a team including those in the pediatric and urologic spheres. However, the report is presented in the hope that consideration will be given to the possible use of muscle transportation procedures in the management of the major abdominal defects occurring in PBS. A personal communication from the president of the Prune Belly Syndrome Network, now on the Internet (http://www.prunebelly.org), indicates that the muscle transposition procedure is unknown to him, and to those worldwide afflicted by the anomaly. It is also possible that it is unknown to their physicians. Finally, extensive experience with transposing thigh muscles shows the procedure to be a reliable procedure with minimal complications and sequela (Ger, in press), the benefits clearly outweighing the risks.

REFERENCES


**Fig. 1.** Protuberant abdominal wall of 13-year-old patient with PBS before muscle transposition surgery. The left costo-vertebral incision resulting from left renal surgery. (Lateral view, and the ventral incision in the anterior view (B) are seen.

**Fig. 2.** Diagrammatic illustration of transportation of the right rectus femoris (R) and left sartorius (S) muscles. The blood supply of the former is by the ascending and transverse branches of the lateral circumflex artery, the descending branch being divided (*) to increase mobilization. Multiple small incisions of the posterior aponeurotic surface of the rectus femoris are seen (arrows). The sartorius is supplied by the superficial circumflex branch of the femoral and ascending branch of the lateral circumflex arteries. For purposes of clarity, one muscle on each side is illustrated.

**Fig. 3.** Anterior (A) and lateral (B) views 3 weeks after surgery. The protuberance shows obvious improvement.
**Fig. 4.** Eighteen years postoperatively. On the lateral view (B), the contour of the left rectus femoris muscle can be seen (arrow). The lines of the incisions are visible on the anterior view (A).

**Fig. 5.** Patient holding the sit-up position using his transposed muscles.
Video Interviews with Dr. Ger and Frank Walker

Below are a series of videos between Dr. Ger and Frank Walker describing Prune Belly Syndrome and Muscle Transposition Surgery:

http://www.youtube.com/watch?v=dvfbBspdJSY&feature=related
http://www.youtube.com/watch?v=tdvm6zjrHY&feature=related
http://www.youtube.com/watch?v=ymbarWwQnjk&feature=related
http://www.youtube.com/watch?v=reG90NHXb3s&feature=related
http://www.youtube.com/watch?v=dLUpVzCXIts&feature=related
http://www.youtube.com/watch?v=aQJZkNJfVfA&feature=related
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