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Management of the Chest-Wall Deformity in Male Patients with Poland's Syndrome

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The chest wall deformity associated with PS was reconstructed in eight male patients 16 to 38 years old (average age 20 years). Follow-up ranged from 1 to 10 years. Two patients had custom silicone implants placed subcutaneously. In one of these patients, the edge of the implant could be seen. Three patients had transfer of an ipsilateral pedicled latissimus dorsi muscle flap with intact thorocodorsal nerve. All these patients had noticible atryphy of the flap, and one underwent subsequent implantation of a custom silicone implant beneath the flap. Three other patients had a custom silicone implant covered immediately by a litissimus dorsi muscle flap. All four patients who had a combination of silicone implant and latissimus dorsi muscle flap had satisfactory correction of their deformity.

Reconstruction of the chest wall in male patients with PS is chellenging because one is attempting to achieve symmetry with a normal contralateral side. Over the last 10 years, we have treated eight male patients with chest-wall deformities attributed to PS. Several observations made in our management of the first few patients led to the refinements used in our current treatment of this deformity.

Patients and Methods

Since 1980, we have treated 29 patients with chest-wall deformities related to PS, 8 of whom were male. These eight patients ranged in age from 16 to 38 years, the average age being 20 years. Follow-up has ranged from 1 to 10 years. All but one of these patients had a deficiency in the subcutaneous tissues of the chest wall. Three patients underwent correction of the deformity by transfer of an ipsilaateral pedicled latissimus dorsi muscle flap with intact thoracodorsal nerve. Two patients had placement of custom silicone implants fabricated from a chest moulage. Three patients underwent placement of custom silicone implants that were immediately covered with a latissimus dorsi muscle flap. One of the patients initially treated with a latissimus dorsi flap subsequently had placement of a cistom silicone implant beneath the muscle flaps at a second procedure. The insertion of the latissimus dorsi was sutured beneath the insertion of the clavicular head of the pectoralis major to reconstruct the anterior axillary folds in four of the six patients in whom the latissimus muscle was transferred.

Case Reports

Case 1

A 21-year-old mas presented with a severe chest-wall deformity characterized by aplasia of the sternal head of the pectoralis muscle; a retrodisplacement of the third, fourth, and fifth ribs of the costochondral junctions anteriorly; and a cephalad displacement of the nipple-areola complex. There was a moderate deficiency of subcutaneous tissues invloving the chest wall and an ipsilateral hand deformity characterized by ulnar angulation of the distal phalanx of the middle finger.

Through an axillary incision, the latissimus dorsi muscle was elevated and transposed by means of an anxillary tunnel to the right anterior chest wall. A preformed cutom silicone implant was inserted beneath the latissimus dorsi muscle. The patient did extremely well, but one year later he developed a sudden hemotoma that required reexploration. No active bleeding was seen, but there was a tear in the cephalic aspect of the capsule. The hemotoma was

evacuated, and the prosthesis and latissimus dorsi flap were left in place. Three years later the result remains excellent.

Case 2

A 38-year-old man presented with a moderate chest-wall deformity related to PS. The deformity was characterized by aplasia of the sternal head of the pectoralis major muscle with a moderate subcutaneous tissue deficiency. The patient underwent placement of a custom silicone implant that was covered by an innervated latissimus dorsi muscle flap. Eighteen months later, the aesthetic result is satisfactory.

Case 3

A 16-year-old boy presented with a moderately severe chest-wall deformity characterized by aplasia of the sternal head of the pectoralis muscle, subcutaneous tissue deficiency, and a depression of the central portions of the second through fourth ribs. The patient was treated with a latissimus dorsi flap, but within 1 year, the muscle had atrophied to the extent that a residual contour deformity could be seen. A custom silicone implant was fashioned and inserted through the original midaxillary incision to establish a satisfactory symmetry with the opposite side. Four and one-half years later, the aesthetic result is satisfactory.

Results

Both patients who underwent placement of custom silicone implants alone had a satisfactory correction of the contour deficiency. One patient was overweight and had adequate soft-tissue coverage of the implant. In the other patient, the outline of the implant was evident on careful inspection.

Of the three patienmts who underwent correction of the deformity with latissimus dorsi flap transfer alone, only one patient had a completely satisfactory correction of his chest-wall deformity. This patient had had a very mild deformity. The other two patients had had undercorrection after atrophy of the latissimus dorsi. One of these patients (case 3) subsequently underwent secondary placement of a custom silicone implant beneath the ttansferred latissimus dorsi muscle. The other patient declined further durgery. The combination of of transferring the latissimus dorsi muscle and inserting a custom silicone implant in three patiets who had more severe deformities achieved a much more satisfactory correction of contour. All four of the patients with a silicone implant beneath the latissimus dorsi muscle were pleased with their chest-wall contour.

One patient required needle aspiration of a serous collection 10 and 17 days postoperatively. Complications in two other patients required secondary operations. One of the patients, in whom only a silicone implant had been placed, developed severe postoperative pain in the area of the implant. The implant was eventually removed, and a second implant was inserted 1 year later withoput any undue problems. A second patient who had had an implant and a latissimus muscle transfer (case 1) developed a sudden hematoma beneath the latissimus dorsi muscle 1 year after operation.

Discussion

PS is an anomoly characterized by thoracic wall abnormalities, vertebral anomalies, and defortmities of the upper extremity. The thoracic wall deformity was first described by Lallemand in 1826. In 1841, Alfred Poland reported the postmortem evaluation of a patient with deficient pectoralis major muscle and syndactyly of the hand on the ipsilateral side. The syndrome was named Poland's syndrome by Clarkson in 1962, after he had operated on a case similar to that described by Poland more than a century earlier.

The thoracic wall abnormalities in PS include deficiency of the pectoralis major muscle, abnormality of the costal cartilages and anterior portion of the ribs, especially the second through fourth ribs, and variable deficiency of the latissimus dorsi, deltoid, supraspinatus, and infraspinatus muscles. The abnormalities also may include aplasia or hypoplasia of the breasts and often the overlying nipple-areola area. We also have noted a deficiency in the amount of chest-wall subcutaneous tissue in almost all the male patients. Most affected women, in contrast, have sufficient body fat for this subcutaneous deficiency to be obvious only in the more severe cases.

The deformity is seen in 1 in every 25,000 births. It is sporadic in most cases, although familial patterns of inheritance have been reported. The ration of male-to-female patients is 3:1, and for unknown reasons, the deformity involves the right side in 75 percent of patients. The most common upper extremity deformities include syndactyly and brachydactyly. Many patients demonstrate hypoplasia of the extremity, which may range from hypoplasia of a single digit to total agenesis of the arm. Other associated anomolies include scoliosis, Sprengel's deformity, dextrocardia, pectus excavatum, renal hypoplasia, foot anomolies, congenital spherocytosis, and an increased incidence of leukemia.

Hester and Bostwick first described latissimus dorsi transposition for the correction of the male Poland's chest-walldeformity. Haller et al. described reconstruction of the chest-wall deformity with an autologous rib graft covered with a latissimus dorsi muscle flap. Seyfer et al. also presented a series that included eight male patients, and their impression was that latissimus dorsi muscle transposition is the procedure of choice in mild chest-wall deformities, whereas more severe deformities require sternal reconstruction with repositioning of cartilage followed at a second stage by latissimus dorsi muscle transposition. Our impression, based on our early experience, is that thepectoralis muscle is bulky muscle in most male patients. Transfer of the latissimus dorsi muscle gives an excellent correction in the initial months following transfer, but once the muscle has atrophied, it is inadequate for all except the very mildest of deformities. If the patient has more than just a simple hypoplasia of the muscle and has a true aplasia of at least the sternal hjead – as is the case in the majority of patients with PS – the latissimus dorsi muscle alone will not be adequate. Whereas autologous rib grafts represent a reeasonable approach for the more severe deformities, custom silicone implants also will adequately camouflage the deformity resulting from absent or malpositioned ribs, and the procedure has the advantage of being very simple with minimal morbidity.

It was our initial impression that the contour deformity in both moderate and severe cases could be corrected just with a silicone implant alone. With increased experience, we concluded that because of the relative deficiency of subcutaneous tissue in many of these patients, there was inadequate soft tissue to camouflage the underlying prosthesis. This conclusion has led us to cover the prosthesis with a latissimus dorsi muscle flap in most cases. The patient may appear overcorrected initially, but once the muscle flap atrophies, an excellent contour symmetry can be established. It is important to transfer the insertion of the latissimus dorsi muscle in male patients. In the one patient in whom we failed to do so, there was a noticeable fullness in the area where the latissimus dorsi was tunneled. With the relative deficiency of subcutaneous tissue, a marked contrast between the fullness of the tunnel and the area below with its paucity of soft tissue became quite noticeable. In those patients in whom the insertion of the muscle was transferred, a natural-appearing anterior axillary fold was created.

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Discussion
by Alan E. Seyfer, MD

It is a pleasure to discuss this article by Marks et al. on the treatment of Poland's syndrome in male patients. Poland's anomaly is indeed an interesting constellation of congenital deficiencies of unknown etiology.

In its mildest form, there is partial or total absence of the pectoralis major muscle; in its severest configuration, there may be absence of ribs, scoliosis, mammary aplasia, and absence of the latissimus dorsi, serratus anterior, and other muscle units of the affected hemithorax and upper extremity. It is of interest that with the full-blown complex, the entire hemithorax is somewhat shortened with a low-riding clavicular head cartilage agenesis and malformation,

abnormally high insertion of the rectus abdominus muscle, and paradoxical motion of the thinned chest wall over the area of rib agenesis. this area characteristically has only a thin element of skin and pleurofascial membrane separating the lung from the external environment .

In our recent review of 55 chest-wall reconstructions for Poland's anomaly, our experience with custom-made, soft-silicone chest-wall prostheses was dismal. Despite the presence of the velour backings and tabs that had been sutured to the clavicular periosteum, three of four patients necessitated removal of the implants owing to adverse cosmesis, migration, and impending exposure. This led us to abandon this mode of treatment and to favor use of the latissimus dorsi muscle, folded upon itself, for morte natural fullness using autogenous tissue. In the female, the simple submuscular insertion of a standard silicone gel mammary prosthesis also gave excellent results and precluded the use of custom-made chest-wall prostheses. This, of course, differs from the experience in the present article. For meaningful comparisons, it is helpful to distinguish the mildest form of the deformity from the more severe form.

Our experience with 21 women indicates that many who receive an initial mammary prosthesis desire no further chest-wall reconstruction, although some may request areolar restoration later. Those who have a more severe deficiency may require latissimus transfer to restore the natural fullness in the subclavicular region. A submuscular mammary prosthesis then fills out the lower portions of the breast.

Unresolved issues include the problem of the axillary foldand the dilemma one faces when there is *absence of the latissimus dorsi* muscle yet the patient requires an autogenous tissue transfer. The axillary fold may be ameliorated by transferring the insertion of the latissimus dorsi muscle. However, these is usually some residual deficiency, and for those of us who perpetually seek improvement in results, its is obvious that the axillary fold is still very different after this maneuver when compared with the normal side. Regarding the absence of a latissimus dorsi muscle, it is hard to justify the use of a superiorly based rectus abdominis musculocutaneous flap (with the skin deepithelialized for more soft-tissue fill) in an adolescent patient. For this limited indication, my inclination would be to use a chest-wall prosthesis in the male patient and a mammary prosthesis in the female patient. These authors' first patient is of interest because he is a "mix" between the severe form (with cartilaginous deformity, low clavicular head, and rib asymmetry) and the mild form (absence of the sternal. head of the pectoralis major muscle.) Again, this brings up the fact that Poland's anomaly is a spectrum of deformities and that the surgical options for cosmetic improvement should be individualized.

It is well to remember that if one looks closely enough, it is possible to detect subtle defects on the affected side of the chest wall. these changes, of course, are still there despite cosmetic improvement, and this fact tends to place any potential goals into perspective. There is little associated disability in this syndrome, even in the most severe configuration. If there is a problem, it is usually related to an upper extremity or shoulder girdle weakness.

Over the years, I ave noted an increase in the number of articles on Poland's anomaly. It is possible that with an increased awareness of this syndrome among pediatricians, family practitioners, and other surgeons, there will be more operations aimed at correcting this cosmetic problem. Hopefully, the plastic surgeon will be able to select from a wide array of alternatives while keeping the reconstruction as simple as possible.

As a parting shot, I feel that there is no place for fasciae latae grafts, methyl methacrylate, or rib-graft reconstructions of the chest wall. These latter procedures have usually ended in failure.

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