SURGICAL TREATMENT OF PATIENTS WITH POLAND’S SYNDROME – OWN EXPERIENCE

MARTA FIJAŁKOWSKA, BOGUSŁAW ANTOSZEWSKI

Department of Plastic, Reconstructive and Aesthetic Surgery Second Chair of Surgery, Medical University in Łódź
p.o. Kierownika: dr hab. B. Antoszewski

Poland’s syndrome is characterized by variable clinical picture. Classic deformity consists of unilateral hypoplasia or aplasia of the sternocostal head of the pectoralis major muscle and ipsilateral hand malformations. Additionally, on the affected side hypoplasia or aplasia of the breast in women is observed. Variable malformations in Poland’s syndrome cause that there is no uniform treatment schedule.

The aim of the study was to analyze surgical treatment conducted in patients with Poland’s syndrome in Plastic, Reconstructive and Aesthetic Surgery Clinic.

Material and methods. A retrospective analysis of surgical treatment was carried out in the group of 66 patients with Poland’s syndrome on the basis of their medical documentation.

Results. The most common operation performed in women was breast reconstruction with silicone implant. Two men had their chest symmetrized by fat grafting. Syndactyly observed in patients with complete Poland’s syndrome was surgically removed in early childhood.

Conclusions. Treatment of patients with Poland’s syndrome, mainly due to variable clinical picture, is individual and depends on age, sex and degree of deformity. In children with complete Poland’s syndrome operations include surgical removing of syndactyly. Breast reconstruction in women with Poland’s syndrome with silicone implants is known and safe method. Such procedures are performed after 18 years of age.

Key words: Poland’s syndrome, hand’s malformations, surgical treatment, breast underdevelopment

Poland’s syndrome was first described by the British surgeon Alfred Poland in 1841. It is characterised by a variable clinical picture, while all the potential symptoms are rarely present in a single patient. The typical components of this syndrome include unilateral hypoplasia or absence of the sternocostal portion of pectoralis major muscle and hand defects affecting the same side of the body (1-4). In females with Poland’s syndrome, impaired development of the breast is observed on the affected body side. Breast growth inhibition has many forms, from mild micromastia to amastia (1, 3-6). In female patients with Poland’s syndrome, the affected nipple-areola complex is usually smaller and less pigmented than the one on the healthy side of the body or may be completely absent (atelia) (1, 4, 6, 7). It is believed that Poland’s syndrome is the most common cause of unilateral congenital micromastia or amastia in females (6, 8).

Coexisting defects of the chest wall and the hand are referred to as complete or full-blown Poland’s syndrome (1, 6). However, more commonly, patients suffer from isolated pectoralis major muscle hypoplasia or aplasia and are diagnosed with partial Poland’s syndrome (1, 4, 6). Many authors agree that the key, constant and sufficient symptom allowing the Poland’s syndrome diagnosis are pectoralis major muscle abnormalities (6, 9, 10).

Due to the large number of defects encompassed by Poland’s syndrome, there has been no uniform treatment regimen established to date. The therapeutic approach is chosen individually for each patient, depending on the severity of pathology, patient’s age and gender (1, 4). In patients with complete Poland’s syndrome, hand surgery is performed first. As
plastic surgery, syndactyly corrections are mainly performed. Occasionally, advanced abnormalities require the amputation of individual non-functional fingers and/or hand prosthesis application (11, 12). Unfortunately, certain forms of defects remain outside the surgical treatment reach (1, 11). The second group of procedures performed in Poland’s syndrome are chest surgeries. Achieving symmetry in patients with Poland’s syndrome remains a significant challenge even for an experienced surgeon (13). The degree of difficulty of the procedure depends on the type of chest deformation. A marked rib hypoplasia or aplasia and coexisting sternal or vertebral defects cause paradoxical chest movements and do not provide adequate protection for the heart and lungs (1, 14). In such cases, procedures stabilising the chest skeleton, performed by thoracic surgeons, are required. More common indications for surgery are aesthetic reasons and psychological aspects, and such surgeries are performed after the patient goes through the developmental stage of life, i.e. reaches the age of 18 (1, 5, 8, 14, 15). The objectives of such surgeries include the filling of infraclavicular fossa and formation of anterior axillary fold (16, 17). In females with Poland’s syndrome, an important part of treatment is the creation of breast on the affected side, matching in its shape and size the breast on the healthy side; of importance is to make symmetrical the whole breast or just the nipple-areola complex (8).

There are a large number of surgical methods applied in the treatment of Poland’s syndrome, and the choice of the technique is associated mainly with the defect advancement stage and the surgical centre preferences.

The aim of the study was the analysis of surgical treatment methods used in patients with Poland’s syndrome at the Department of Plastic, Reconstructive and Aesthetic Surgery, Medical University in Łódź.

MATERIAL AND METHODS

In the years 1990-2010, 66 patients with Poland’s syndrome were treated at the Department of Plastics, Reconstructive and Aesthetic Surgery, Medical University in Łódź. Clinical examination was performed and photographs taken in all the patients. Based on the medical documentation (medical history, outpatient sheet, photographs), a database of patients with Poland’s syndrome was created. The collected data included patient’s age, gender, affected side of body, distinction between complete and partial defect sets, and surgical treatment methods used.

A retrospective analysis of surgical treatment methods used in patients with Poland’s syndrome was performed based on the patient medical documentation and the created database. The surgical procedures performed in patients were divided into two groups. In the first one, breast reconstruction methods in females and chest wall symmetrisation in males were under analysis, while in the second one – hand surgeries in children with complete Poland’s syndrome.

RESULTS

Females constituted the majority of patients with Poland’s syndrome treated at the Department (58 individuals – 87.9%, as opposed to 8 males – 12.1%). The reason for the higher ratio of females reporting for consultation and treatment was breast asymmetry causing them stress, caused by micromastia on the affected body side. The analysed patients were aged between 6 and 65, with the mean age standing at 28.22. Right-sided defect set was present in 31 patients (47%), while left-sided one in 35 (53%). Complete Poland’s syndrome was diagnosed in 19 patients (28.8%; 13 females and 6 males), while the remaining 47 individuals had diagnosed partial Poland’s syndrome (71.2%; 45 females and 2 males).

In females, the most frequently performed breast reconstruction surgery on the affected side was silicone implant insertion (47 patients, 79.3%) (fig. 1). Implant size was selected individually depending on the breast size on the healthy side; the mean implant volume was 235 cm³ (from 175 cm³ to 300 cm³). In 9 patients from this group, due to the breast sagging and/or breast size on the healthy side, procedures aimed at breast symmetrisation (reduction and/or lifting of the healthy breast) were performed (fig. 2). Two patients (3.45%), due to the great advancement of the defect (amastia on the affected side), were first qualified for expander implantation. Upon filling the expander with saline to the size matching that of the breast on the healthy side, the second part of the procedure was performed.
expander replacement with a silicone breast implant. Two females (3.45%) were qualified for breast reduction and lifting on the healthy side. The performed procedure allowed to achieve a symmetrical breast in relation to the hypoplastic breast and the final aesthetic result was fully satisfactory for the patients. In 2 patients, additional surgical procedures were performed aimed at symmetrisation of the nipple-areola complex. In one patient, nipple-areola complex reduction was performed on the healthy body side, while in the second one – nipple and areola enlargement on the affected side. The remaining 7 female patients were consulted at the Plastic Surgery Clinic at young age (paediatric patients) and they will undergo qualification for breast symmetrisation upon reaching the age of 18.

The group of males with Poland’s syndrome treated at the Department encompassed 8 patients, of which 6 suffered from complete Poland’s syndrome and 2 – partial. In 1 patient with partial Poland’s syndrome, chest wall asymmetry was slight and there were no definite indications for surgical intervention. In 2 patients, marked pectoralis major muscle hypoplasia on the affected side caused a visible disproportion in the chest wall anatomy. The above defect was stressful for the patients to a degree justifying qualification for surgical treatment.

Autologous fat grafting (from buttocks) was performed in them, with a satisfactory aesthetic result. In one boy, marked chest wall asymmetry was observed, requiring skeletal correction, and the patient was referred to the thoracic surgery centre. The remaining patients were consulted at young age (paediatric patients) and they will undergo potential qualification for chest symmetrisation procedures upon reaching the age of 18.

In the group of patients with complete Poland’s syndrome, 8 individuals (5 females and 3 males) were qualified for surgical syndactyly treatment by digital separation. All procedures were performed under general anaesthesia, at limb ischaemia. Syndactylic digit separation was accompanied by web formation with the use of either 2 triangular flaps or 1
rectangular flap (fig. 3). In 4 patients, syndactyly affected fingers 2-5 and in those cases, separations of fingers 2 and 3 and fingers 4 and 5 were performed at one time. In no procedure a finger was separated from the others on both its sides to prevent blood supply impairment. Syndactyly of fingers 3 and 4 was treated at the next stage. Six patients required surgical enlargement of finger web. Syndactyly digit separation was performed at kindergarden age (between the ages of 2 and 6) (fig. 4). Only 1 female patient underwent the procedures at an older age (9 and 10) due to the fact that patient’s parents had not reported to the hospital until she was 9. Upon the treatment completion, patients were able to perform alternating finger movements, impossible previously, and two of them started learning how to play keyboard instruments.

The remaining 11 patients with complete Poland’s syndrome had no indications for surgical treatment of the upper limb. In 7 of them, the hand on the affected side was formed correctly, although smaller than the healthy one. In one female patient, the abnormal hand was smaller that the healthy one and in addition fingers 2 and 3 were deformed, while two other patients were diagnosed with ectrodactyly (lobster-claw syndrome). In one patient from this group, the hand defect was so advanced (absence of fingers 2-5) that it required consultation at the hand defects centre.

DISCUSSION

Polymorphism of the clinical picture of Poland’s syndrome has precluded the development of a uniform treatment regimen in patients with this complaint. The therapeutic approach depends mainly on the degree of pathology severity, patient’s age and gender. Surgical procedures performed in patients with Poland’s syndrome fall into two categories: upper limb surgeries and chest repair procedures.

Hand defects were diagnosed in 19 patients with Poland’s syndrome among those treated at the Department of Plastic, Reconstructive and Aesthetic Surgery, and 8 of them were

Fig. 3. A scheme of syndactyly release using 2 trigonal flaps (A) and a rectangular flap (B) to create a interdigital web; the defects on lateral sides of the fingers are covered with free full thickness skin grafts

Fig. 4. A patient with complete right-sided Poland’s syndrome before and after surgical release of syndactyly

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qualified for surgical procedures. Surgery consisted in the separation of simple syndactyly. In patients with syndactyly of fingers 2-5, separations of fingers 2 and 3 and fingers 4 and 5 were performed at one time. In no procedure a finger was separated from the others on both its sides, and syndactyly of fingers 3 and 4 was performed at a later stage. Such an approach prevents blood supply impairment and is in line with published reports (4). Surgical separation of syndactylic digits was performed in paediatric patients at the age of 2-6. Authors are in agreement that such age is optimal for this type of procedure (4, 11, 12). It is important to start the treatment early in order to improve the hand function. Of significance is also the fact that surgical separation of syndactyly may involve several stages, and if initiated at proper age allows the treatment completion before the child goes to school and starts learning how to write. More advanced upper limb defects require treatment at the hand surgery department or orthopaedics department. Unfortunately, certain deformations remain outside the available therapeutic potential (1, 11).

The second category of procedures performed in Poland’s syndrome encompasses chest surgeries. In females, such procedures are aimed at the reduction of asymmetry in the breast size, while in males – in the chest wall. In the studied group of patients, the most frequently performed surgery in females with Poland’s syndrome, where the primary defect was micromastia while the pectoralis major muscle defect caused slight asymmetry in the chest wall, was the enlargement of affected breast by means of a silicone implant. Implant selection was individual, depending on the healthy breast size. In some females, due to the size and sagging of the breast on the healthy body side, it was decided to perform its lifting and/or reduction. In patients with amastia, expander was inserted at the first stage of treatment, followed by its replacement with a breast implant. A similar therapeutic approach has been recommended by Ribeiro and colleagues; in most severe forms of the defect the authors suggest using dermomyocutaneous flaps (18). Many other authors also consider breast implants as the method of choice in the treatment of female patients with Poland’s syndrome (1, 5, 15, 19). There have also been reports published offering criticism of silicone implants and recommending the use of dermomyocutaneous flaps (1, 20). However, it should be borne in mind that silicone implant insertion is a relatively low invasive procedure. The use of dermomyocutaneous flaps is associated with the formation of additional scars and the weakening of back or abdominal muscles. Breast implants are most commonly inserted from under the breast, owing to which the scar is hidden in the inframammary fold and the achieved aesthetic result is judged as good or very good.

In the studied group of patients, breast reconstruction procedures were performed in individuals aged 18 or above. At that time, the breast on the healthy body side reached its relatively final size and allowed correct implant selection. The authors of other publications are in agreement as to the timing of performing this type of procedure; breast symmetrisation is also recommended not before the patient reaches the age of 18 (1, 5, 14). In males with chest asymmetry stemming from pectoralis major muscle aplasia, without coexisting skeletal defects, the indications for the procedure include mainly aesthetic reasons. In 2 patients with Poland’s syndrome treated at the Department, aesthetic surgery was performed involving fat grafting into the cavity. Such a type of procedure is a recognised method of tissue filling and is recommended by some authors also in patients with Poland’s syndrome (21, 22). Some surgeons use in males with Poland’s syndrome individual prostheses mimicking the pectoralis major muscle (23). Unfortunately, this type of implants is made to order by specialised companies only and greatly increases the costs of the procedure.

CONCLUSIONS

1. The treatment of patients with Poland’s syndrome, due to the variable clinical picture, is selected individually and depends on patient’s age, gender and pathology severity.
2. The treatment of patients with complete Poland’s syndrome is started at the kindergarden age, and the surgeries involve the separation of syndactylic digits.
3. Breast reconstruction with the use of silicone implants in patients with Poland’s syndrome is a recognised and safe method of forming a breast, providing a good aesthetic result. Such procedures are performed in patients aged 18 and over.
REFERENCES


Received: 15.11.2011 r.
Adress correspondence: 90-1533 Łódź, ul. Kopcińskiego 22