Treatment of congenital elevation of the scapula

10 (2–18) year follow-up of 37 cases of Sprengel's deformity

Bernhard Greitemann, Jan-Jomit Rondhuis and Alfred Karbowski

We studied retrospectively and partially prospectively all 37 cases of Sprengel's disease who were treated between 1970 and 1990, 23 of whom were operated on. Many of the patients were severely handicapped by abnormal shoulder abduction and elevation. Neurological deficits were not found. 34 of our cases had other congenital malformations.

Several operative techniques were used. In cases with only cosmetic problems we now prefer resection of part of the superior angle. In cases with impaired function we prefer the Woodward-procedure.

[1]Klinik für Techn. Orthopädie u. Rehabilitation and [2]Klinik für Allgemeine Orthopädie, Westfälische Wilhelms Universität, Robert Koch Strasse 30, D-4400 Münster, Germany. Tel +49–251 83 6767 or 6761. Fax –251 83 6776
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With improved radiographic techniques more cases of Sprengel's deformity are diagnosed (Laumann and Cire 1985). The deformity is not only characterized by elevation of the scapula. An adduction and medial shift of the scapula, a distal rotation with tilting of the glenoid cavity, a more or less disturbing prominence of the superior angle of the scapula and changes in the position of the clavicula are also found (Jeannopoulos 1961, Cavendish 1972, Laumann and Cire 1985). We report our experience with Sprengel's deformity over a 20-year period.

Patients and methods

We analyzed 37 patients with Sprengel's deformity (congenital elevation of the scapula) who were treated in our department during 1970 and 1990. All cases with Klippel-Feil-syndrome were excluded. Patients were examined retrospectively and partially prospectively by clinical and radiographic examinations, recently also by stereophotogrammetry. All patients were followed for at least 2 years postoperatively with a mean of 10 (2–18) years. 10 patients were female and 27 male. The median age when first seen was 5 years (3 months–18 years).

Many of the patients were severely handicapped; 14 reported problems in everyday life, 27 in sports, and 6 in their work. The cosmetic problems were felt very severely by 2 patients, severely by 11, slightly by 9, and not even slightly by 16. Those who had severe or very severe cosmetic problems were worried more by these than by their shoulder function (Figure 1).

Functional deficits affected mainly abduction and elevation of the arm, as well as external rotation. Clinically, elevation showed a mean of 123° vs 177° on the non-affected side, and abduction a mean of 128° vs 175°. Elevation of the scapula was measured radiographically as the difference between the affected and non-affected sides in the inferior angle of the scapula. The mean difference was 3.4 (1–7.5) cm. The medial shift of the scapula was measured by the distance between the medial margin of the scapula and a vertical line from the spinous process of C7. The mean distance was 3.8 cm on the affected side and 6.8 cm on the non-affected side. The mean distal rotation was 3° by stereophotogrammetry.

Neurological deficits were not found. The movements of the head were not affected, but 6 patients had

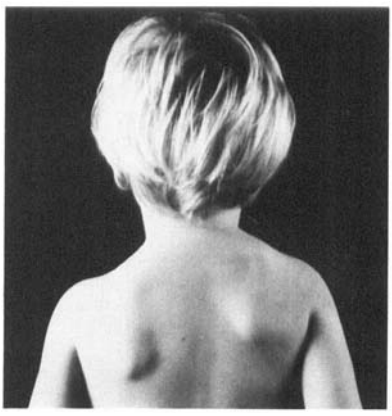

Figure 1. Typical feature of Sprengel's disease.

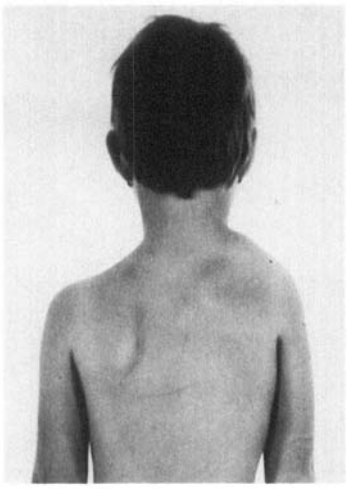

Figure 2. Cavendish 3 and 4 deformity.

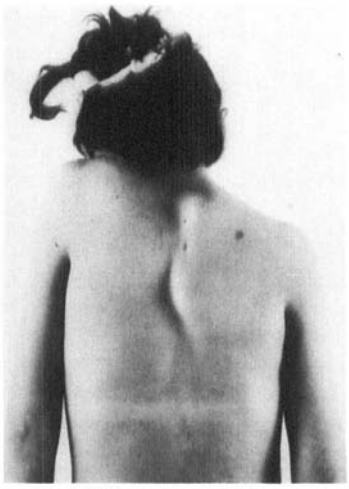

Figure 3. Os omovertebrale.

asymmetrical posture of the head. 7 patients complained of pain in the arm or cervicothoracic spine (6 patients) and the surrounding muscles. 1 patient had chronic headache. 9 patients had atrophy of the rhomboid, the trapezoid, and the pectoral muscles. 2 patients had facial asymmetries.

In evaluation of the clinical severity of the disease, we used the Cavendish (1972) classification which is mainly focused on cosmetic aspects. 18 patients had severe forms of scapular elevation, i.e., Cavendish 3 or 4 (Figure 2).

The Cavendish classification was correlated with the radiographic examination and the clinical measurements of function and therefore gave an acceptable classification of the deformity (Table 1).

In 14 patients a connection between the superior border of the scapulae and the cervical spine was found, not always bone, but sometimes fibrous tissue. A real omovertebral bone, referred to as a rudimentary rib by Putti (1908), was found in 7 patients (Figure 3). In 34 of our 37 patients we found other congenital diseases, notably bone deformities, ear deformity, double kidneys, vessel anomalies, pulmonary disorders, hip dislocation, foot deformities, etc.

Table 1. Correlation clinical function/Cavendish classification

Mean elevation of the shoulder	Cavendish Type	n
153°	1	10
133°	2	9
104°	3	15
90°	4	3

Deformities of the ribs were found in 30 patients with synostosis, too few or too many ribs, or changes in shape or figuration. 18 of these patients had scoliosis with Cobb angles of more than 20°. The main curve was located in the cervicothoracic and upper thoracic spine in 11 of those patients. In 13 patients the curve was located on the side of the deformity, and in 5 cases on the opposite side.

Operations

23 patients were treated operatively. The indications for operation were deficits in shoulder movements, usually abduction of less than 120°, or severe psychological problems caused by the cosmetic appearance.

The choice of operation was based on the Cavendish classification. Patients with deformity 1 or 2 only, were treated by resection of the prominent part of the superior angle of the scapula. This is only a cosmetic repair; the malrotation of the scapula and the functional deficits in shoulder movements are not corrected. Patients with deformity 3 or 4 were treated by distalization of the scapula. Here the main muscles of the scapula also are transposed, and most of the deformity is corrected.

During the 20-year period the operative procedures used for severe cases changed. In the early years the König-Wittek procedure (König 1913) was done in 10 patients with separation of the muscles from the scapula, osteotomy of the scapula and distalization of the lateral part. Because of scar-keloid, difficulties in osteotomy and bone irregularities postoperatively, as well as poor functional improvement, this procedure was abandoned, and the Green (1972) procedure was done in 2 cases. This procedure also caused difficulties with

Table 2. Mean operative results (n 23)

	All	Resect.	Distaliz.
Reduction of scapular elevation, cm	2.5	1.8	2.7
Improvement in Cavendish Type	0.7	0.2	0.85
Improved elevation, degrees	19	5	23

scar-keloid, and there was another change to the Woodward (1961) procedure which was used since 1981 in 6 patients. 5 patients had only a resection of the disturbing part of the supraspinatus portion of the scapula and of an omovertebral bone. The mean age of the operated patients was 8 (3–17) years.

The mean elevation in the operated group was 4.1 cm preoperatively and 1.6 cm postoperatively (Table 2). The patients who had an operation with distalization of the scapula had a mean reduction of elevation of 2.7 cm, and those with resection the superior angle of the scapula had a reduction of 1.8 cm.

The mean improvement in the Cavendish classification was 0.7, and the improvement in function was a better elevation and mean abduction by 19°. There was no correlation between age and results.

We found the best results with Woodward's procedure. 4 of 5 patients showed good results, the mean improvement in function was 38°, and the mean cosmetic improvement was 1.3 classes. 4 of the 5 patients classified as good had undergone a Woodward procedure with a mean reduction of elevation of 3.5 cm (Figure 4).

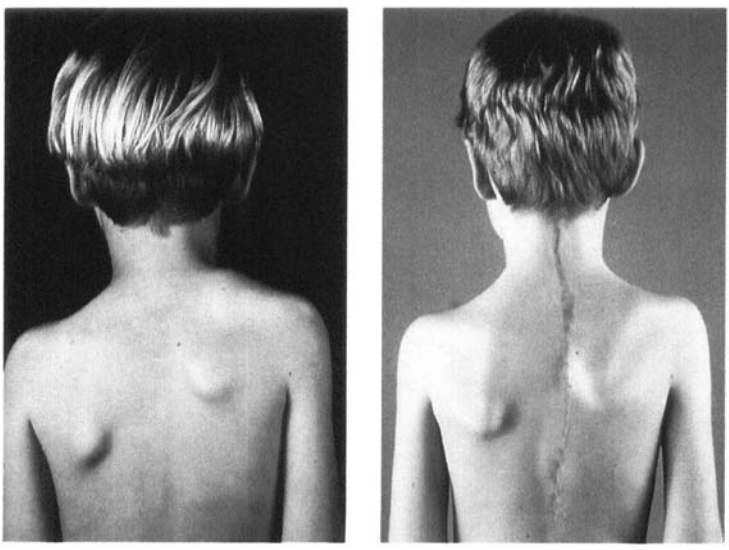

Figure 4. Patient with Cavendish 2/3 deformity preoperatively and after Woodward's procedure.

8 patients had complications. There were 4 cases of scar-keloid, all after Green and König procedures. 3 patients had osteophytes after a König procedure. 1 case of Woodward operation had a transient plexus paralysis.

Discussion

Sprengel's disease is a rare malformation and therefore no large series has been described. Laumann and Cire (1985) made a comprehensive review of all published cases.

In our analysis of operations we found the Woodward procedure to be the one with the best results. The average increase in elevation of the arm reported in the literature ranges from 23° to 57° (Robinson 1926, Wilkinson and Campbell 1980, Laumann and Cire 1985). The result is surely dependent on the population of patients. Therefore, the mean effect on elevation in our patients may seem rather small. But we think that there are several reasons for this. One is the strict selection of patients. The transposition procedures were only done in cases with severe functional deficits. The other operative procedures improve only the cosmetic appearance, not the function. Therefore, this group lowers the overall result.

There are several reasons for better results with the Woodward procedure. First, there is a lower risk of forming a scar-keloid which may fix the scapula in a poor position, because the muscles are incised further from the scapula. Second, a larger mobilization is possible. Third, the postoperative scar is not so thick as, for instance, in Green's procedure. But this procedure runs the risk of damaging the plexus—it cannot be controlled intraoperatively (Woodward 1961). An osteotomy of the clavicle may be performed to prevent such complications. This has now become our routine procedure.

In our opinion the indication for operative treatment should be very strict. It seems reasonable to do only a resection of the superior angle of the scapula if there are only cosmetic problems and no functional deficits. If there are deficits in function, then a Woodward procedure should be done, with careful examination of the plexus postoperatively, possibly

with electromyographic control intraoperatively. We very much favor an osteotomy of the clavicula if greater distalization is desired.

Although we could not find any correlation of the operative result with the patients' age, we agree with others (Jeannopoulos 1961, Woodward 1961, Wilkinson and Campbell 1980, Laumann and Cire 1985) in proposing that the best age for operation is under 6 years, when the soft tissues are more flexible and a better correction can be achieved. Every child with Sprengel's deformity should be examined carefully for associated defects; these confirm the prenatal cause of the deformity.

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