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CONGENITAL HIGH SCAPULA

(Sprengels Deformity)

By

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Congenital high scapula ("Elevation of the scapula") is an uncommon deformity, in which the shoulder girdle, which develops as a cervical appendage, fails to descend properly during early foetal life. This condition was first described by *Eulenburg* (1862) but it became better known through *Sprengel*'s report of 1891, its commonest denomination originating from this report. The causes of this deformity are obscure and various suggestions have been put forward with regard to the therapy.

We have found no mention of Sprengel's deformity in the Scandinavian literature. At the meeting of the Finnish Surgical Association in 1925 Professor R. Faltin demonstrated a bilateral case as a curiosity. We therefore consider it justified to present four cases treated at the Children's Clinic.

The nature and symptoms and other simultaneous deformities appear from table 1 and from the figures. The superior border of the scapula was in two cases four, in one case three and in one case two vertebrae too high.

INCIDENCE, ETIOLOGY AND PATHOGENESIS

That Sprengel's deformity is a rare condition is proved by the fact that in 1904 *Erhardt* collected 92 cases from the literature and that in 1942 *Junge* arrived at the figure of 250 cases. The proportion of bilateral and unilateral cases is 1: 10 (*Ombrédanne*). *Junge* (1942) reports an incidence of 1 among 2–3000 other congenital orthopedic deformities.

During the period 1947 to 1953 about 3300 different congenital deformities were treated in the surgical department of the Children's

TABLE 1

Essential data for 4 operated cases of Sprengel's deformity.

Case record	Sex	Age years	Side	Age at which deformity was noted	Symptoms	Other developmental disturbances observed
1 4081 1952	F	3	Right	6 months	No abduction above the frontal plane. Omovertebral bone.	Lumbosacral meningocele. Costal asymmetry and deformity.
2 591 1952	M	4	Left	At birth	No abduction above the frontal plane. Locking of shoulder joint. Pain. Short clavicle.	Vertebral scoliosis. Synostosis of the 7th cervical and 2nd thoracic vertebra. Spina bifida occulta of the 6th cervical vertebra.
3 1414 1953	F	2	Right	1 year	Normal motility.	Vertebral scoliosis. Costal deformity. The 3rd thoracic vertebra incomplete.
4 4081 1953	F	7	Left	At birth	Abduction 50° above the frontal plane. Hook-shaped projection on the scapula.	Synostosis of the 3rd cervical and 3rd thoracic vertebra. Fissure of the hard and the soft palates. Congenital ptosis of the right upper eyelid. Hyperopia.

Fig. 1.
Case 1. 3-year-old girl with congenital high scapula.

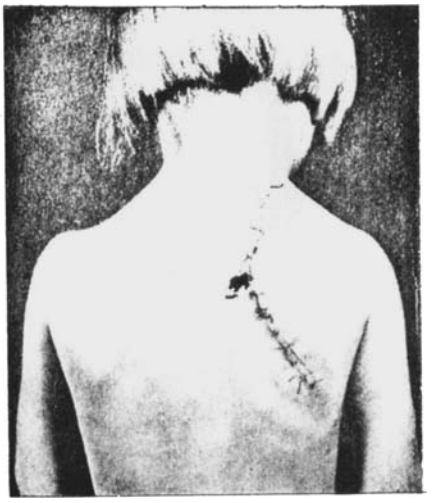

Fig. 2.
Case 1 after operation.

Clinic. Since this material was selected in many different ways, Sprengel's deformity is apparently no commoner in Finland than elsewhere in the world. We estimate that one such patient is born in Finland every fourth year.

Erhardt (1904) and *Cohn* (1907) distinguish between a congenital and an acquired form. The difference is obscure. According to them the great majority of cases are, however, congenital. *Inclán* (1949) divides the cases into two groups depending on whether there are spinal changes or not. This division is of practical importance in the prognosis.

Sprengel's deformity is generally considered a partial failure of the upper extremity to descend during foetal life. The first sac-like anlage of the upper extremity appears in the fourth foetal week and from it the different parts of the upper extremity then gradually develop. In the beginning the anlage of the scapula is situated at the level of the fourth cervical to the second thoracic vertebra, descending during the second and third foetal month, simultaneously with an initial ossification of the scapula, in a caudal direction to the level of the second to eighth rib. Since the clavicle on the side of the deformity has often proved shorter than normal, it has been assumed that in the descent the scapula is not able to follow the clavicle but remains somehow riding on the shoulder (*Ombrédanne*).

Numerous other theories besides these have been presented with regard to the etiology and pathogenesis of the condition. Sprengel as-

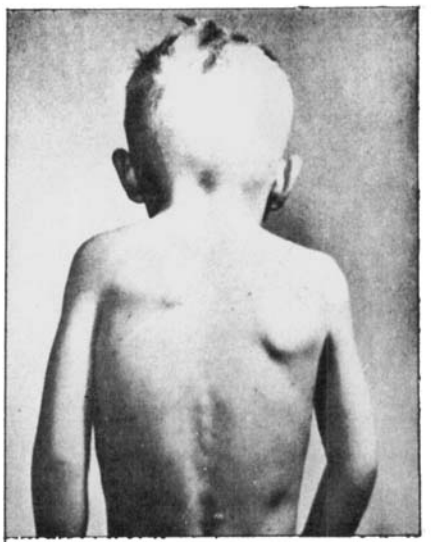

Fig. 3.
Case 2. 4-year-old boy with operated Sprengel's deformity (plus Klippel-Feil's syndrome).

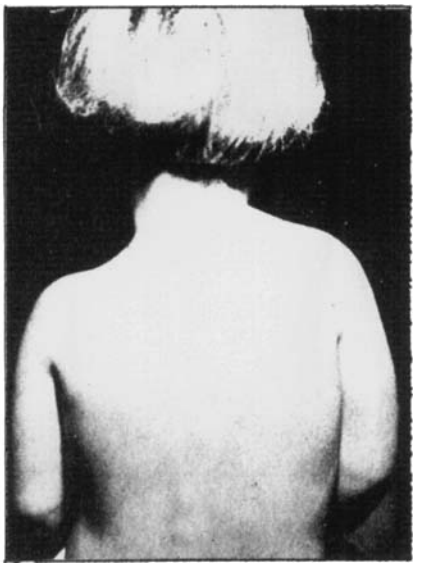

Fig. 4.
Case 3. 2-year-old girl with congenital high scapula.

sumed the cause to be upward pressure of the uterine wall due to too little amniotic fluid. However, cases in connection with hydramnion have been reported, and moreover this theory in no way explains the other deformities of which, according to *Junge* (1942), there is always a minimum of one and in 50 per cent of cases several, accompanying Sprengel's deformity (Figs. 7, 8, 9).

Because of the vertebral and costal deformities which are the commonest deformities associated with Sprengel's deformity, many authors consider a faulty mesodermal segmentation the primary cause (*Cohn, Putti, Fairbank, Teske, Inclán*). Rachischisis formations and Klippel-Feil's syndrome in association with Sprengel's deformity have sometimes been explained, on the lines of Engel's (1943) bleb theory, as being the result of leakage of the cerebrospinal fluid into differentiating tissues in the fifth and sixth foetal weeks.

In the recent literature (*Keith,* 1933, and *Ingersoll,* 1945) congenital high scapula has been explained as an atavism because in some instances the deformity occurred together with an omovertebral bone which prevents descent and which is found in certain species of fish and frogs. The series collected by *Horwith* (1908) included 27 such cases and later *Carson* (1938) and *Rösger & Ebert* (1940) each described one case. In our series (case 1) an omovertebral bone was

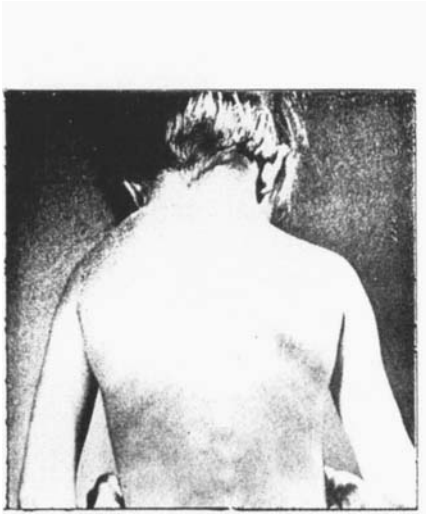

Fig. 5.
Case 4. 7-year-old girl with congenital high scapula (plus Klippel-Feil's syndrome).

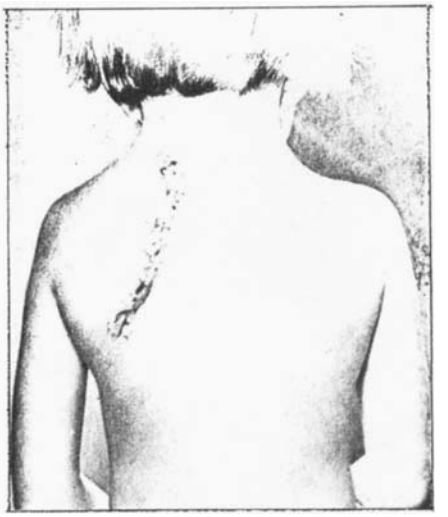

Fig. 6.
Case 4 after operation.

roentgenologically established (Fig. 8). At operation a 1.5 cm. wide bony bridge articulating with the scapula and attached to the spinous processes with fibrous tissue was found. Further the same patient had costal deformities and lumbar meningocele, so that the case can no more be considered an atavism than it can be explained by the bleb theory alone.

Neuhof (1913) and *Engel* (1939) have found deformities in relatives of their patients, although heredity is generally not invoked.

In our cases the simultaneous frequency of other deformities is striking. The most natural explanation of deformities of such varying type and location in so many different segments and organs would be early injury to the fetus by some noxa. The presence of an omovertebral bone (case 1) need not be a separate phenomenon but an early disturbance of the ontogenetic development may result in phylogenetic development forms.

TREATMENT AND RESULTS

In Sprengel's deformity the diagnosis is as a rule obvious. From a differential diagnostic point of view thoracic asymmetries, paralysis of the anterior serratus muscle, sequels of polio, hysteria, muscular dystrophy, spondylitis and neoplasma should, however, be remembered.

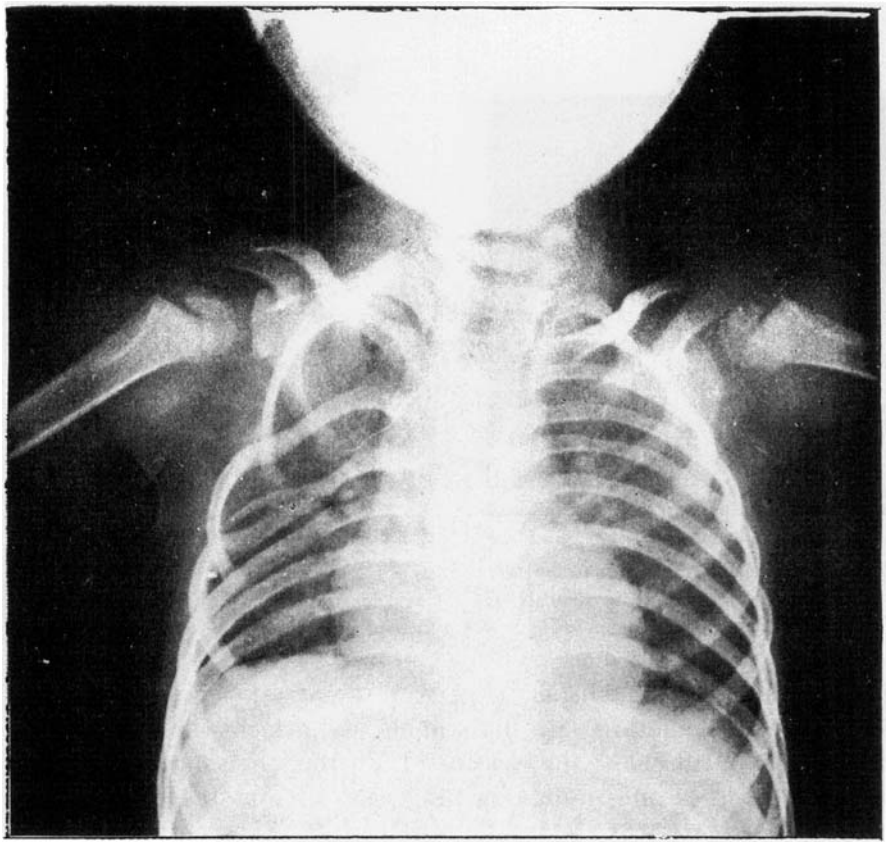

Fig. 7.
Roentgenogram of case 3 before operation.

In our cases the indication for treatment, apart from external asymmetry, was limited abduction of the upper extremity in three cases and severe pain in the shoulder in one case.

Of the many operative methods described in the literature we have principally followed that described by *R. D. Schrock* (1926) which resembles the technique related by *Putti* almost 20 years earlier. Operative method: A skin incision is made along the vertebral border of the scapula. The rhomboideus, anterior serratus and subscapularis muscles are subperiosteally dissected. The subscapularis muscle is dissected totally free. The infraspinatus and the teres major and minor muscles are freed so that the caudal angle and two thirds of the caudal border are entirely exposed. The trapezius, supraspinatus and levator scapulae muscles are further dissected subperiosteally. Dissection at the cranial angle must be cautious in order to avoid injury to the nerves. Any fibrous or chondro-osseous bridge from the spine to the

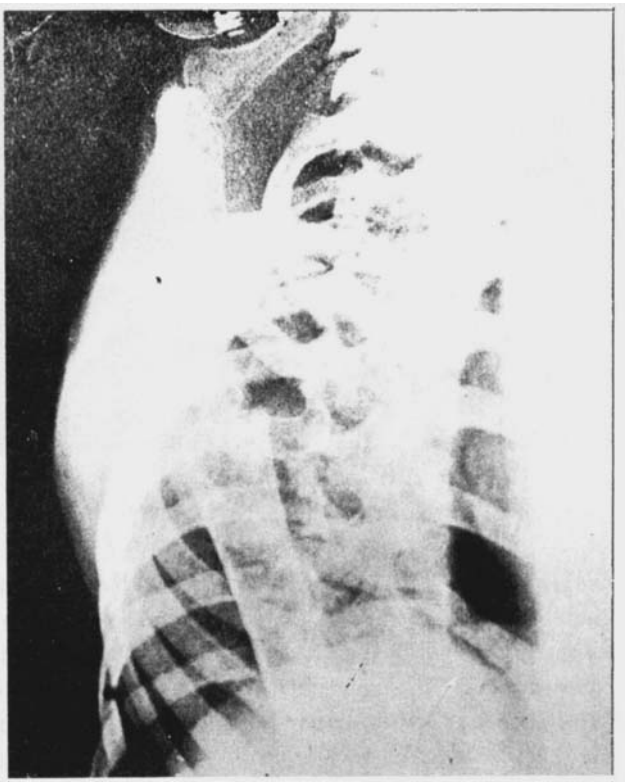

Fig. 8.
Free projection of the scapula in case 1 shows an omovertebral bone articulating with the cranial angle of the scapula.

scapula is thoroughly extirpated. The vertebral border of the scapula is resected so that the scapular index, which in Sprengel's deformity is high, is reduced. Then redressement of the shoulder region and rotation of the scapula in the shoulder joint in a caudal direction is made (Fig. 11).

By means of metal wire the scapula is fixed to the lowest possible rib. Exact closure in layers with silk is made. An elastic bandage is applied over the shoulder. We have not used plaster casts.

No special after-treatment was given but our patients have been allowed to exercise the shoulder joint freely when playing.

The postoperative course was uneventful. In case 3 we found immediately postoperatively that the metal attachment had broken but the result was nevertheless satisfactory and reoperation was therefore not performed.

A complication mentioned in the literature is that either the brachial plexus or the brachial artery may become compressed on

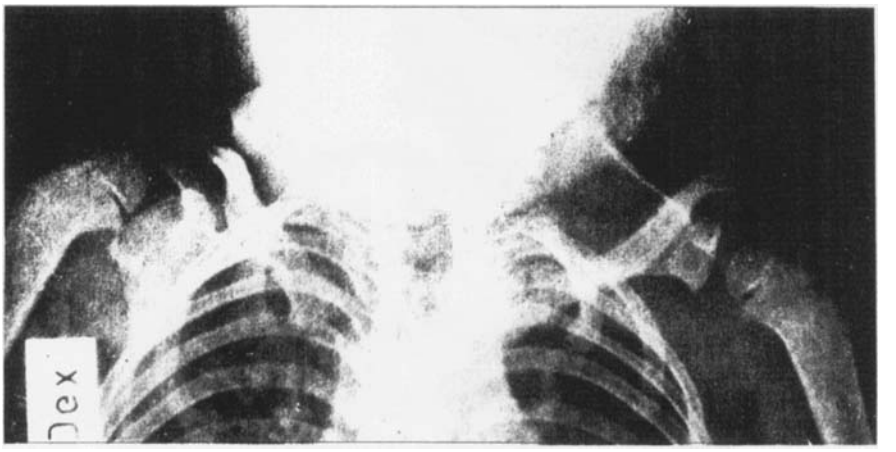

Fig. 9.
Roentgenogram of case 2 before operation.

descent of the shoulder. This is avoided if at redressement the radial artery is watched and if after anesthesia the motility and sensitiveness of the extremity are checked. If the descent of the shoulder is not sufficient osteotomy of the acromion or the clavicle is recommended. We did not find these additional measures necessary in our cases.

By this technique we achieved good balance with regard to the scapula in all our cases. In case 3 there was a partial recurrence. The external cosmetic deformity improved considerably in all cases and in those cases in which the abduction of the arm had been limited, the motility became normal. The pain in case 1 disappeared.

At after-examination one year or more after operation the primary operative result persisted, or had slightly improved. Calcifications due to the subperiosteal dissection were slight and of no importance (Fig. 10).

As compared to the healthy shoulder the operated side was 2, 2.5, 2 and 0.5 cm. higher. This asymmetry was in all cases hidden by the clothes.

Operative treatment of a scapula that has failed to descend will lead to a perfect result only in those rare cases in which the thorax and the vertebrae are quite normal. In all our cases there were other simultaneous deformities affecting the external cosmetic result. The outcome was in all cases, however, good since the relative invalidity of the patient was considerably reduced.

In the literature four years is given as the minimum age of operated cases of Sprengel's deformity. Of our cases three had just reached four years or were younger. As far as the operative risk is concerned, opera-

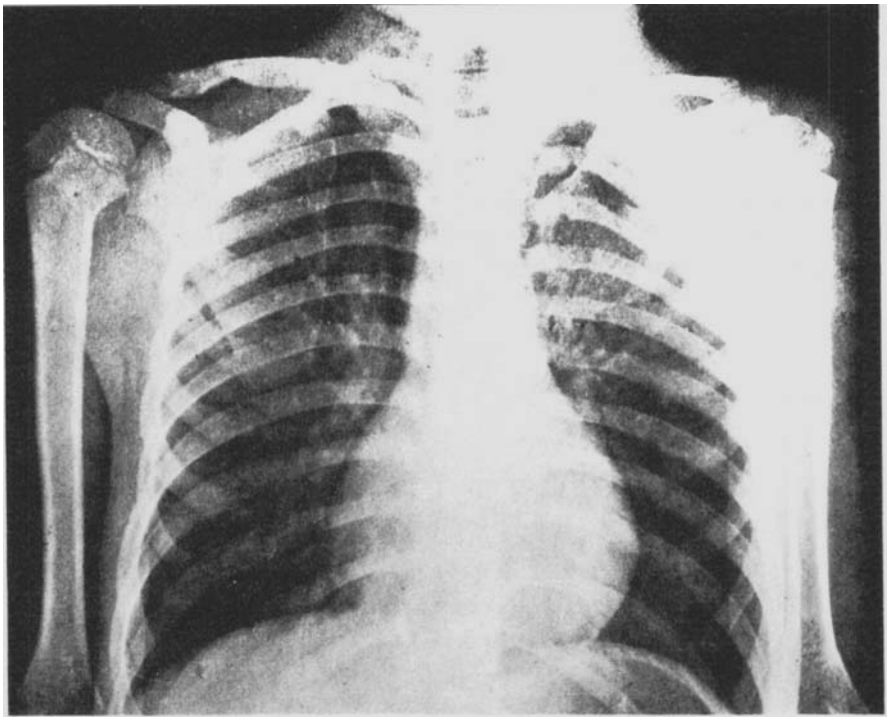

Fig. 10.
Roentgenogram of case 2 after operation.

tions of this type can be performed at any time after the first year of life. Bearing in mind secondary scolioses and contracture positions we feel inclined to operate as early as possible after the first year. As appears from our cases little co-operation is required of the patient in any kind of after-treatment, good motility being obtained when the scapula is rotated into the normal position.

SUMMARY

The authors present 4 cases of Sprengel's deformity at the age of two to seven years. The patients exhibited simultaneous spinal deformities, two patients costal deformities, one meningocele and one uranostaphyloschisis. Cosmetic asymmetry and in three cases limited abduction and in one pain in the shoulder were the operative indications. The patients were operated on principally according to Putti-Schrock's method. The authors describe their technique. Operation revealed that the scapula on the deformed side was attached by fibrous adhesions to the spine, whilst the scapula itself exhibited abnormal

processes. In one case there was a real omovertebral bone (Fig. 8). After subperiosteal dissection of its vertebral portion, the scapula is brought down in a caudal direction by rotation in the shoulder joint (Fig. 11). Fixation is best made to the costae.

The cosmetic result was good in three cases and satisfactory in one. Abduction of the arm above the frontal plane became quite normal in all cases. The postoperative course was uneventful. The authors recommend operation at the age of one to two years.

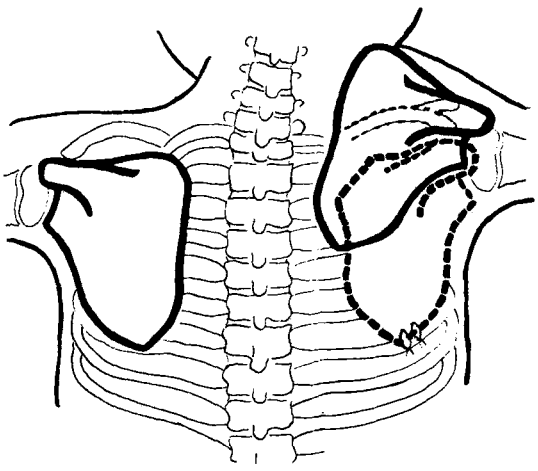

Fig. 11.

Schematic presentation of congenital high scapula. The hatched area shows the position of the scapula after correction and caudal rotation in the humero-scapular joint. The caudal angle of the scapula resected.

RESUME

Les auteurs présentent 4 cas de déformité Sprengel à l'âge de 2 à 7 ans. Les malades avaient simultanément des déformités rachidiennes, deux malades des déformités congénitales, un méningocèle et un autre uranostaphyloschisis. Une assymétrie cosmétique avec abduction limitée dans trois cas et des douleurs de l'épaule dans l'autre ont été les indications de l'opération. Les malades ont été opérés principalement d'après la méthode Putti-Shrock. Les auteurs décrivent leur technique. L'opération a révélé que du côté déformé, l'épaule était rattachée à la colonne vertébrale par des adhésions fibreuses et qu'elle présentait elle-même des processus anormaux. Dans un cas il y avait un os directement omovertébral. Après dissection subpériostale de la partie vertébrale, l'épaule est abaissée en direction caudale par rapport à l'articulation. La meilleure fixation se fait sur la côte.

Le résultat cosmétique a été bon dans trois cas et satisfaisant dans un. L'abduction du bras au-dessus du plan frontal est redevenue entièrement normale dans tous les cas. Le cours postopératoire n'a présenté aucun caractère particulier. Les auteurs recommandent l'opération à la âge de un à deux ans.

ZUSAMMENFASSUNG

Die Verfasser berichten über 4 Fälle von Sprengels Missbildung im Alter von zwei bis sieben Jahren. Die Patienten wiesen gleichzeitig Wirbelsäulenmissbildungen, zwei Patienten Rippenmissbildungen, einer Meningocele und einer Uranostaphyloschisis auf. Verunstaltende Assymetrie, in drei Fällen eingeschränkte Abduktion und in einem Schmerzen in der Schulter Liessen die Operation angezeigt erscheinen. Die Patienten wurden im wesentlichen nach der Putti-Schrock Methode operiert. Die Verfasser beschreiben ihre Technik. Die Operation offenbarte, dass das Schulterblatt der missbildeten Seite mit fibrösen Verwachsungen an die Wirbelsäule fixiert war, während die Scapula selbst abnorme Gestaltung zeigte. In einem Falle war ein richtiger omovertebraler Knochen vorhanden. Nach subperiostaler Freilegung ihrer vertebralen Portion wird die Scapula in caudaler Richtung mittels Rotation im Schultergelenk heruntergebracht. Es ist am besten, sie an einer Rippe zu fixieren.

Das kosmetische Ergebnis war gut in drei und zufriedenstellend in einem Falle. Die Abduktion des Armes wurde normal in allen Fällen. Der postoperative Verlauf war ungestört. Die Verfasser empfehlen die Operation im Alter von ein bis zwei Jahren.

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