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The Sprengel deformity

MORPHOMETRIC ANALYSIS USING 3D-CT AND ITS CLINICAL RELEVANCE

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We evaluated scapular dysplasia and malposition in 15 patients with the Sprengel deformity using three-dimensional CT (3D-CT). The shape, height-to-width ratio, the areas of both scapulae, the anterior curvature of the supraspinous portion and glenoid version were assessed on scapular posterior, medial and inferior views. The degree of rotation and superior displacement were measured on the trunk posterior view. The omovertebral connection was also assessed and correlated with the operative findings.

Most of the affected scapulae had a characteristic shape with a decrease in the height-to-width ratio and were larger than the contralateral scapulae. There was an inverse relationship between scapular rotation and superior displacement. The typical curve of the supraspinous portion of the scapula was seen in only three cases. There was no significant difference in glenoid version. The point of tethering of the omovertebral connection may determine the shape, rotation and superior displacement of the scapula. 3D-CT was helpful in delineating the deformity in detail, and in planning scapuloplasty.

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The Sprengel deformity is a complex anomaly of the shoulder girdle associated with malposition and dysplasia of the scapula with muscle hypoplasia or atrophy, which causes disfigurement and limitation of movement of the shoulder. Various procedures have been described for the surgical correction of this rare deformity, including simple resection of prominent bone,[1] relocation of the scapula by detaching muscles from their insertion into it[1-3] or by detaching and transplanting them at their midline origins,[4] and by combinations of these procedures.

Most of the anatomical descriptions of patients with the Sprengel deformity have been based on physical examination, surgical exploration, or plain radiographs. The last are of limited value in assessing deformity since the scapulae overlie the trunk, making their bony margins difficult to identify; they are rotated in the coronal, sagittal and horizontal planes. There is only one description of a cadaver specimen with a Sprengel deformity in the English literature.[5]

Three-dimensional CT (3D-CT) has been found to be useful in defining complex anatomical structures.[6,7] We have therefore assessed the Sprengel deformity by this method in order to clarify the anatomical abnormalities and to plan surgical treatment.

Patients and Methods

Between September 1994 and March 1999, we used 3D-CT in 15 patients with the Sprengel deformity. There were nine boys and six girls. In 13 the deformity was unilateral with the left scapula affected in eight and the right in five. In the remaining two, the condition was bilateral with the left side affected more severely. The mean age of the patients was 4 years and 3 months (3 years to 8 years and 11 months). Twelve patients were treated by operation. We also assessed 13 contralateral scapulae for comparison.

Spiral CT was performed using the HiSpeed Advantage System (GE Medical Systems, Milwaukee, Wisconsin) with a scanning technique of 120 kVp and 200 mA. 3D images were reconstructed with a slice thickness of 3 mm at increments of 2 mm by the Advantage Windows 3D Analysis Package release 2.1 (GE Medical Systems). Trunk anterior, trunk posterior, superior trunk outlet and scapular posterior, medial and inferior views were taken. The trunk anterior or posterior view corresponded to the image of the whole shoulder girdle as seen from the front or behind, that of the superior trunk outlet to the image of the upper trunk seen from above, and the scapular posterior view to the *en-face* image of the scapula as seen from behind. The scapular medial view was obtained by rotating the single scapula around the vertical axis, and the scapular inferior view by rotating it around the transverse axis.

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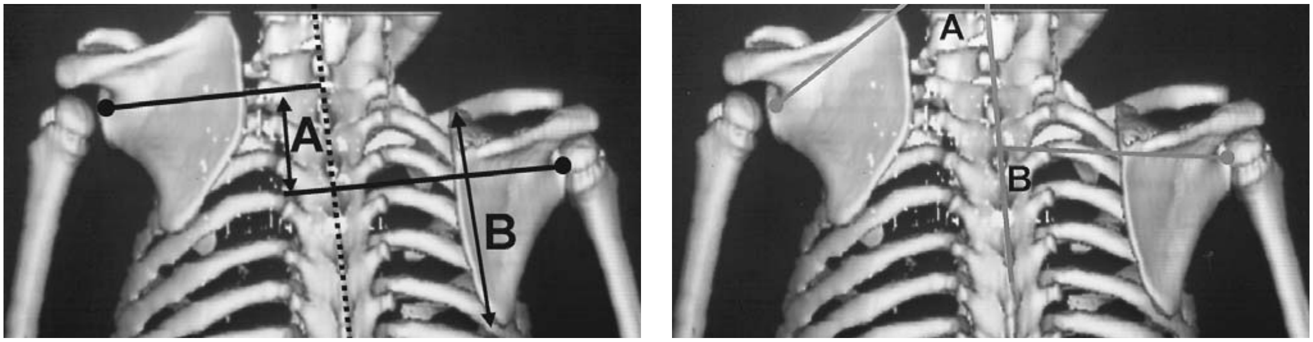

Fig. 1a Fig.1b

Measurement of the superior displacement and the rotational deformity on the trunk posterior view. Figure 1a – Lines are drawn from the centre of the glenoid cavity perpendicular to the vertebral axis line. Superior displacement = A/B. Figure 1b – The rotational deformity of the affected scapula is defined as the difference between the angle of tilt of both sides. Rotational deformity = B-A.

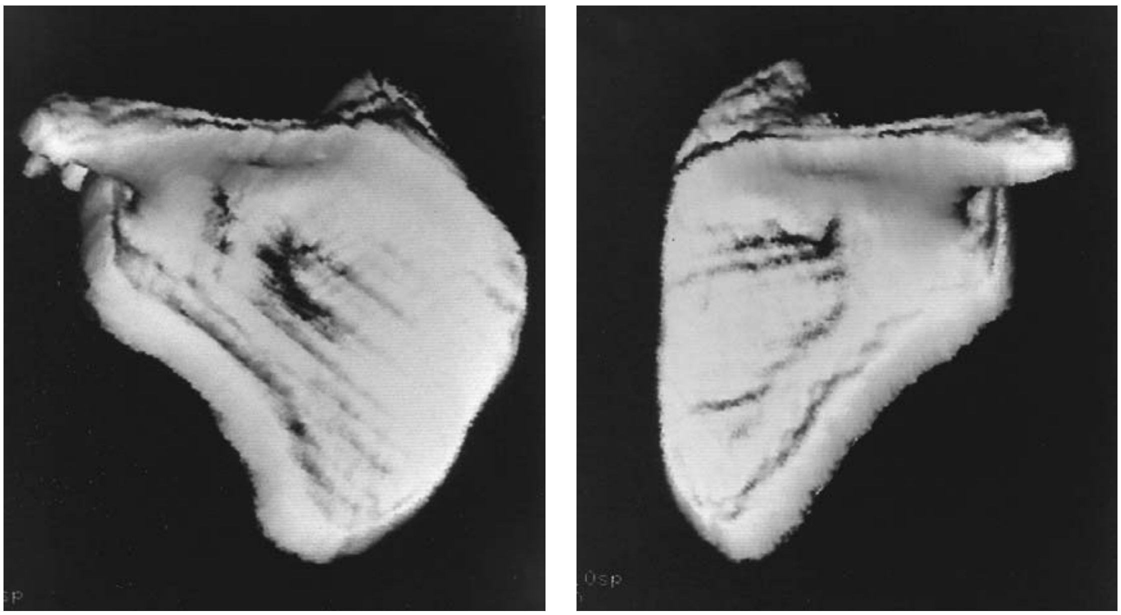

Fig. 2a Fig. 2b

Typical appearance of the Sprengel deformity on the scapular posterior view. The affected left scapula (a) has a convex vertebral border, concave lateral border, and laterally curved inferior angle, and its area is larger than that of the contralateral scapula (b) by 20.2%.

Superior displacement and rotation of the affected scapulae were measured in the unilateral cases on the trunk posterior view with reference to the contralateral side (Fig. 1).

The level of the inferior angle of both scapulae was measured with reference to the thoracic spine on the trunk posterior view.

The scapular contour was evaluated on the scapular posterior view. The affected scapulae were considered to be of typical shape when the vertebral border was convex, the lateral border concave, and the inferior angle curved laterally (Fig. 2). The contralateral scapulae in unilateral cases were graded as normal when the vertebral border was straight along the proximal three-quarters of the infraspinous portion and as borderline when it was mildly convex (Fig. 3). We determined the height-to-width ratio of the scapula on the scapular posterior view. The height was measured from the superior angle to the inferior angle parallel to the cavity of the glenoid and the width from the cavity of the glenoid to the most medial portion of the vertebral border perpendicular to the glenoid. The area of the scapula was determined on the scapular posterior view by computer graphic software (Image-Pro Plus V3.01: Media Cybernetics, Silver Spring, Maryland).

The anterior curvature of the supraspinous portion was evaluated on the scapular medial view (Fig. 4). The glenoid version was measured on the scapular inferior view by a modification of the method of Friedman, Hawthorne and Genez.[8] The bony component of the omovertebral connection and its tethering point to the scapula were evaluated where it was most clearly demonstrated (Fig. 5), and compared with the operative findings. The clavicular con-

Fig. 3a

Fig. 3b

A unilateral case showing a) a normal scapula and b) borderline dysplasia.

Fig. 4a

Fig. 4b

A scapular medial view of a) the contralateral scapula and b) the affected right scapula with definite anterior curving of the supraspinous portion.

tour was assessed on the trunk anterior and superior trunk outlet views and the scapuloclavicular space on the superior trunk outlet view by measuring the angle of convergence of the longitudinal lines of the shaft of the clavicle and the spine of the scapula.

We tested the correlation between superior displacement and the rotational deformity by Spearman's non-parametric correlation test in 13 unilateral cases. The differences in the height-to-width ratio and glenoid version between the affected and contralateral scapulae were determined by the Wilcoxon rank-sum test, and the difference in the area of the affected and contralateral scapulae by the Wilcoxon signed-rank test in the unilateral cases. A p value of less than 0.05 was considered to be statistically significant.

Results

The 3D-CT data for the 15 patients and the clinical results of the 12 who had surgery are summarised in Table I. The mean superior displacement of the affected scapulae, as measured by the difference in the glenoid level divided by the height of the contralateral scapula, was 0.27 (0 to 0.55). The mean rotational deformity of the affected scapulae in the unilateral cases was 27° (5 to 53). The Spearman test showed a statistically significant inverse correlation between superior displacement and rotation ($p = 0.026$). The more cephalic the glenoid was placed the less the scapula was rotated. The level of the inferior angle of the contralateral scapulae ranged from the sixth thoracic vertebra to the eighth, while that of the affected scapulae ranged from the third to the seventh.

All the affected scapulae including bilateral cases had a characteristic shape, except for two unilateral dysplasias. The contralateral scapulae in unilateral cases had a normal shape in five and borderline dysplasia in eight (Fig. 3). The mean height-to-width ratio of the 17 affected scapulae was 1.23 (0.94 to 1.60), and of the 13 contralateral scapulae 1.40 (1.32 to 1.58). The Wilcoxon rank-sum test showed that the ratio was significantly smaller on the affected than on the contralateral side ($p = 0.0012$). The affected scapulae were significantly larger than the contralateral scapulae in the unilateral cases ($p = 0.014$). The mean area of the

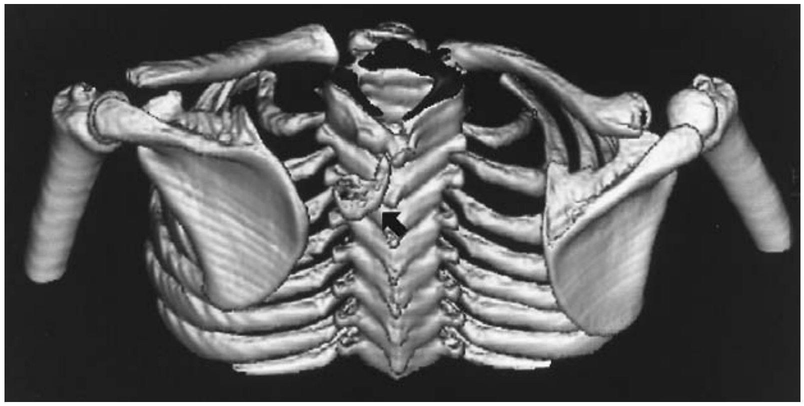

Fig. 5

3D-CT of a bony omovertebral connection.

affected scapulae was larger than that of the contralateral scapulae by 13.6% (-6 to 27) in the unilateral cases (Fig. 2).

Anterior curving of the supraspinous portion was definite in three cases, borderline in seven and less than the contralateral side in five. The mean glenoid retroversion was 0.7° on the affected side and 0.8° on the contralateral side with no significant difference ($p = 0.52$). The affected clavicle contoured slightly differently: the lateral aspect had a greater curvature on the trunk anterior view, and the clavicular shaft appeared to have less curvature forming a narrower scapuloclavicular space on the superior trunk outlet view.

The bony omovertebral connections in eight cases were evaluated by 3D-CT, and faced towards the convexity or prominence of the vertebral border (Fig. 5). In six of the 12 patients who had surgical treatment, it was seen preoperatively on 3D-CT. It corresponded to the operative findings in terms of location, direction and points of tethering to the scapula. Of six cases in which no bony components were visualised on preoperative 3D-CT, four had cartilaginous and two fibrous connections. The omovertebral connections were tethered to the convexity of the infraspinous portion of the vertebral border in all but two of the patients undergoing surgical treatment.

The affected scapulae of these two cases (cases 1 and 6) had an atypical shape. In one (case 1) the vertebral and lateral borders were unusually straight, and the superior angle was elongated resulting in a height-to-width ratio larger than that of the contralateral side. A cartilaginous omovertebral connection was found at operation, which arose from the upper cervical spine, and tethered to the elongated superior angle. The glenoid level of the affected scapula was much higher than the contralateral side, but the rotational deformity was minimal (Fig. 6). In the other case (case 6), the affected scapula had an irregular contour with multiple exostosis-like lesions, and no bony or cartilaginous omovertebral connections were found apart from multiple fibrous bands. The affected scapula showed marked superior displacement with moderate rotation.

As shown in Table I, on the basis of the classification of Cavendish[9] and of measurements of abduction of the shoulder, improvements in cosmesis and function were seen in all patients who had an operation (Table I and Fig. 7).

Discussion

The contours of the scapula are difficult to evaluate by plain radiography because of its position, rotation, and shape.[10] 2D- and 3D-CT scans have been used to evaluate scapular fractures and deformities.[7,8] Malposition and dysplasia of the scapula and the presence, size and tethering points of bony omovertebral connections can be assessed most accurately by 3D-CT.

Several anatomical landmarks such as the level of the shoulder joint,[9] the centre of the scapula,[11] the inferior angle of the scapula,[12] and the centre of the head of the humerus[13] have been used as reference points when assessing malposition. We measured the superior displacement and the rotational deformity separately. The level of the glenoid cavity was used when measuring superior displacement (Fig. 1a) since it represents the connection between the trunk and upper limb and is less affected by rotation. There have been few attempts to measure the rotational deformity. Leibovic, Ehrlich and Zaleske[14] measured the deformity from a line connecting the acromioclavicular joint to the inferior angle of the scapula. We used the base of the spine of the scapula as a reference point for measuring rotational deformity because it was easily identifiable on 3D-CT and was believed to be less affected by dysplasia (Fig. 1b).

The observation of a significant inverse correlation between superior displacement and rotational deformity led to consideration of the role of the omovertebral connection in the pathomechanism of the Sprengel deformity. The omovertebral connection causes limitation of abduction of the shoulder. 3D-CT was sensitive in detecting the bony omovertebral connection. In our study, this was found in more than 50% of cases, which was higher than the 19% to 40% in previous reports,[1,9,13,15] and tethering was usually

Table I. Details of 3D-CT data for the 15 patients and of the clinical results after operation for 12

						3D-CT				Cavendish grade‡		Passive abduction (degrees)			
Case	**Age (yr+mth)**	**Gender**	**Side**	**Scapular shape***	**Omovertebral connection (type, site)**	**Rotational difference† (degrees)**	**Superior displacement ratio†**	**Height-to-width ratio (affected/ contralat)**	**Anterior hooking**	**Preop**	**Postop**	**Preop**	**Postop**	**Surgical procedures§**	**Postop follow-up (mth)**
1	3+8	F	R	Atypical	Cartilaginous, sup. angle	5	0.55	1.60/1.40	Absent	IV	I	110	180	Mod. W	17
2	3+9	F	L	Typical	Bony, infraspinous	32	0.20	1.19/1.34	Borderline	IV	II	90	160	Mod. W	27
3	4+1	F	R	Typical	Bony, infraspinous	48	0.11	1.21/1.46	Definite	IV	I	100	180	Green	38
4	4+9	M	R	Typical	-	45	0.21	1.23/1.37	Borderline	III	-	95	-	None	-
5	4+7	F	L	Typical	Fibrous, infraspinous	16	0.17	0.94	Definite	T_2	T_3	110	160	Mod. W	22
			R	Typical	-	-	-	1.09	-	T_2	T_2	135	180	Excision, sp	22
6	3+1	M	L	Atypical	Bony, multiple irregular	35	0.27	1.44/1.32	Definite	IV	II	120	165	Mod. W	37
7	3+8	M	L	Typical	Cartilaginous, infraspinous	18	0.37	1.23/1.34	Borderline	IV	I	100	160	Mod. W	32
8	3+3	F	L	Typical	Bony, infraspinous	8	0.54	1.10/1.42	Borderline	III	I	110	180	Mod. W	36
9	4+8	M	L	Typical	Bony, infraspinous	33	0.07	1.19/1.58	Borderline	II	-	90	-	None	-
10	8+11	M	L	Typical	Bony, infraspinous	12	0.54	1.16	Absent	C_7	T_1	145	160	Mod. W	10
			R	Typical	-	-	-	1.30	-	T_2	T_2	170	-	None	-
11	3+0	M	R	Typical	Bony, infraspinous	20	0.18	1.21/1.32	Absent	III	-	90	-	None	-
12	3+11	M	R	Typical	Fibrous, infraspinous	53	0.00	1.28/1.44	Borderline	IV	II	120	170	Mod. W	20
13	3+2	M	L	Typical	Bony, infraspinous	14	0.19	1.23/1.37	Absent	III	I	90	170	Mod. W	44
14	5+1	F	L	Typical	Cartilaginous, infraspinous	21	0.43	1.25/1.43	Borderline	IV	II	160	180	Green	62
15	3+8	M	L	Typical	Cartilaginous infraspinous	13	0.35	1.19/1.35	Absent	III	I	90	160	Mod. W	51

* cases 2, 4, 6, 7, 9, 11, 13, 15 showed borderline dysplasia on the contralateral side
† correlation between the superior displacement and rotational deformity was tested only in the unilateral cases
‡ level of the centre of the glenoid was measured in reference to the thoracic vertebral body on the plain anteroposterior radiograph in the bilateral cases
§ supraspinous portion was resected in all operated cases; prominent vertebral border was also excised in cases 3, 5, 8, 10, 15; mod. W, modified Woodward operation; Green, Green operation; sp, supraspinous portion

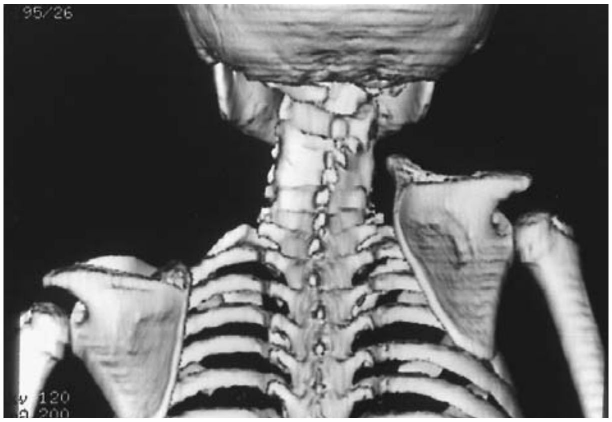

Fig. 6a

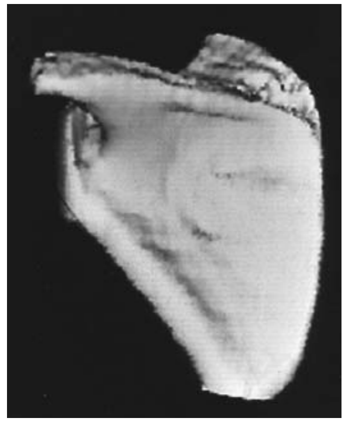

Fig. 6b

Fig. 6c

An atypical case. The cartilaginous omovertebral connection is anchored at the superior angle, resulting in an increased height-to-width ratio. Figure 6a – Trunk posterior view. Figures 6b and 6c – Scapular posterior views.

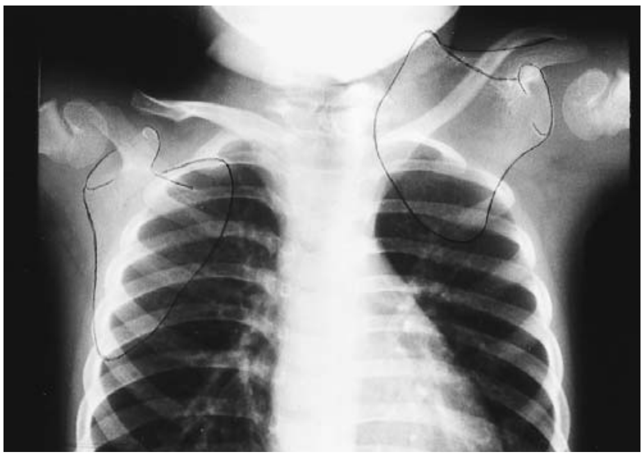

Fig. 7a

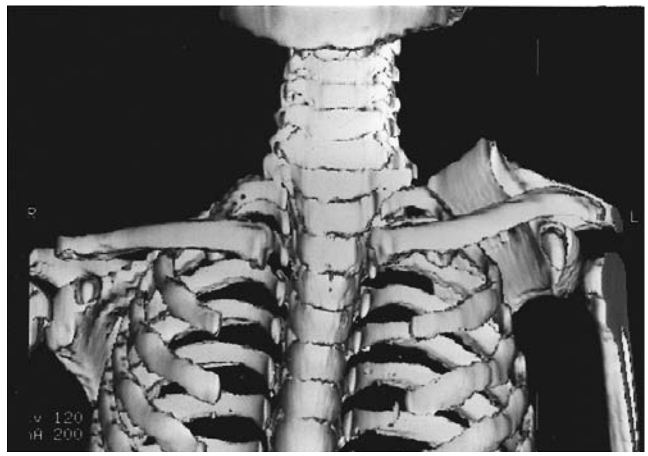

Fig. 7b

Fig. 7c

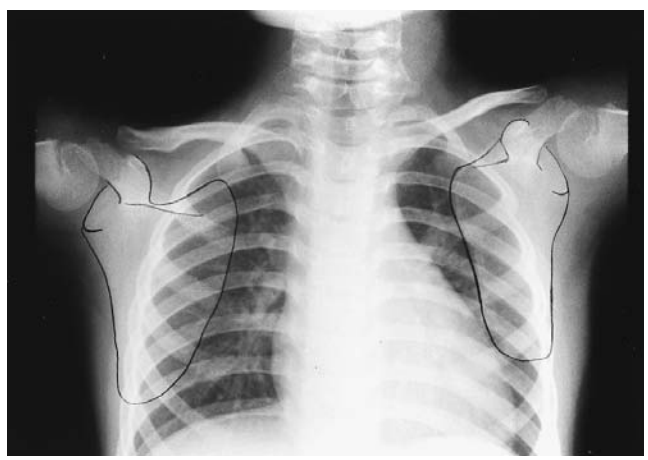

Fig. 7d

Anteroposterior radiograph of the trunk of a three-year-old girl with Cavendish grade-III Sprengel deformity of the left scapula (a). The lateral part of the left clavicle is obliquely elevated and the acromial end has a greater curvature on the trunk anterior view (b), forming a narrower scapuloclavicular space on the superior trunk outlet view (c). An anteroposterior radiograph of the trunk at three years after a modified Woodward operation including resection of the supraspinous portion and excision of the convex vertebral border of the scapula shows definite improvement (d).

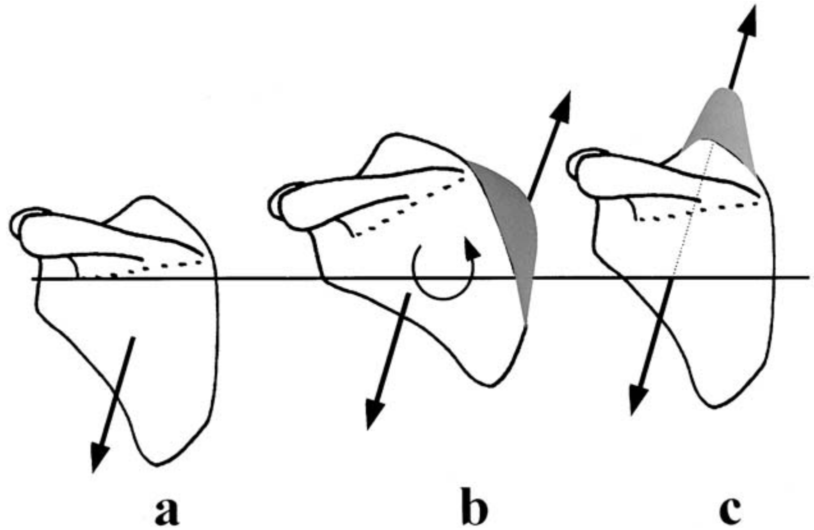

Fig. 8

Illustration of the hypothetical relationship between the tethering structure and malposition. Figure 8a – Normal caudal migration of the scapula during embryonal development. Figure 8b – Tethering medial to the migrating force affected the rotational torque resulting in marked rotation. Figure 8c – Tethering in line with the migrating force left the scapula in the cephalic position with little or no rotation.

at the convexity of the vertebral border of the infraspinous portion of the scapula. In the two cases in which the omovertebral connections were tethered at different points, the shapes and pattern of malposition of the affected scapulae were atypical. The case in which tethering was at the superior angle was at the extreme end of the displacement-rotation spectrum. The omovertebral connection clearly has a major role in determining the shape and malpositioning of the scapula. During embryonic development, it is postulated that a 'pulling force' acts on the scapula in caudal and lateral directions (Fig. 8). If the tethering structure acts in line with this force, the scapula will remain in the cephalic position with little or no rotation, as in one of our atypical cases. If tethering is present medial to the line of the force, however, it will produce a rotational torque. As a result, the scapula will rotate so that the glenoid cavity faces inferiorly, with only a difference in the level of the glenoid cavity. The apparent elevation of the shoulder in this case presented as a fullness at the base of neck caused by the superior angle of the scapula. Most cases of the Sprengel deformity lie in between these two extremes.

The affected scapula is not only malpositioned but also dysplastic. It has been described as hypoplastic,[2,4,11,12,15-17] with a decreased height-to-width ratio,[5,11,12,15,16] and occasionally with an anteriorly curved supraspinous process.[2,11,12,15] There have been, however, few reports of quantitative measurements. Previous descriptions included only radiological assessment with oblique images of the scapula. We measured the area of the scapula on its posterior *en-face* view with the help of computer graphic software, which showed that the affected scapulae were larger than the contralateral scapulae in most cases, and this suggests that the scapular growth was not arrested but aberrant.

There has been only one report of the height-to-width ratio, the mean value of which in 35 normal cadaver scapulae with an age range from 0 to 14 years, was 1.49 (1.42 to 1.56),[18] slightly higher than that of our series. The reasons for this discrepancy may be twofold. The radiolucent cartilaginous portion was included in the measurement while we measured only the osseous contour. Although the cartilaginous contour and its height-to-width ratio change very little during fetal and postnatal development, the osseous contour is invariably smaller longitudinally than laterally.[5,19] The contralateral scapulae in cases of unilateral deformity are occasionally dysplastic with a decreased height-to-width ratio, which suggests that the morphogenesis of the contralateral scapula, even if fully migrated, may be affected.

Ogden et al[5] described a cadaver study in a 14-year-old specimen with unilateral deformity, and later showed the ratio to be 1.04 on the affected and 1.51 on the contralateral side.[18] The ratio of the affected scapulae in our series ranged from 0.94 to 1.60, reflecting the diversity of the pathoanatomy of the Sprengel deformity.

Ogden and Phillips[18] attributed the scapular dysplasia to an alteration in blastemal formation of the pectoral girdle and also to altered lateral growth primarily in the infraspinous portion of the vertebral border physis, which appeared to be related to the omovertebral tethering. This hypothesis is supported by one of our atypical cases, in which the omovertebral connection was tethered at the superior angle instead of at the infraspinous portion of the vertebral border. In this case, the scapular growth was not altered laterally but longitudinally, resulting in a height-to-width ratio higher than that of the contralateral side.

Definite anterior curving of the supraspinous portion was observed in some cases. There was no significant difference in glenoid version between the affected and contralateral scapulae, but it is not possible to conclude that there is no difference in version since the glenoid cavity had not fully ossified in this age group.[18] The affected clavicle contoured differently and our observation is in agreement with Ogden et al[5] in that the acromial end of the clavicle has a greater curvature, and forms a narrower angle with the elevated, forwardly displaced scapula. It was not possible in this 3D-

CT study to evaluate the true dimensions of the costoclavicular space in which the brachial plexus is situated.

The evaluation of the Sprengel deformity by 3D-CT led us to consider whether surgical treatment could be planned according to the pathoanatomy. After releasing the muscular and tethering structures, relocation should focus on the dominant deformity be it superior displacement or rotational deformity. Several authors have recommended that the transplanted scapula should be held by traction through a wire placed in the lateral one-third of the base of the spine of the scapula[2] or at the inferior angle.[20]

Based on the new finding of hyperplasia of the affected scapula, we advocate resection of the prominent convexity of the vertebral border as well as the supraspinous portion, as recommended by Borges et al[15] in selected cases with a markedly decreased height-to-width ratio. The scapular posterior view of 3D-CT was helpful in determining whether or by how much repositioning was possible.

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