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Passive range of glenohumeral motion in children with a Sprengel's deformity

Francisco Soldado, MD, PhD [a,b,*], Pierluigi Di-Felice-Ardente, MD [c], Sergi Barrera-Ochoa, MD, PhD [d], Paula Diaz-Gallardo, MD [a,b], Josep M. Bergua-Domingo, MD [a,b], Jorge Knörr, MD [a,b]

[a] Pediatric Upper Extremity Surgery and Microsurgery, Barcelona Children's Hospital HM Nens, Barcelona, Spain
[b] UCA Unit, Vithas San Jose, Vitoria-Gasteiz, Spain
[c] Shouder Unit, Orthopedic Surgery Service, Hospital Manresa, Manresa, Spain
[d] Hand Surgery Unit, ICATME, Centre Medic Quiron Deixeus, Barcelona, Spain

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Background: In Sprengel's deformity, loss of shoulder motion has been attributed exclusively to scapulothoracic stiffness. The purposes of this study were to evaluate passive glenohumeral (GH) joint motion in these children.

Methods: A prospective evaluation of 23 children was performed. Obtained data were demographics, Cavendish grade, bilateral active global shoulder elevation, and multidirectional passive GH range of motion, including: (a) GH internal rotation in abduction and GH cross-body adduction to assess for posterior GH contracture; (b) spinohumeral abduction angle (SHABD) to assess for inferior GH contracture; (c) spinohumeral adduction angle to assess for superior GH contracture; and (d) passive external rotation in shoulder adduction and abduction to assess for anterior GH contracture. Paired *t* tests and both Pearson's and Spearman's correlation analyses were performed.

Results: The mean patient age was 8.1 years (range, 1.4-16.7 years), with 13.4% of deformities Cavendish grade 1, 52.2% grade 2, 13.4% grade 3, and 21.7% grade 4. The involved shoulder showed a statistically significant decrease in mean active global shoulder elevation (117.4° vs. 176.1°), SHABD (14.6° vs. 41.5°), cross-body adduction (43° vs. 71.3°), and internal rotation in abduction (17.8° vs. 49.4°), all at $P < .001$. Strong inverse correlations were noted between Cavendish grade and both global shoulder elevation (r, −0.784) and SHABD (r, −0.669). Cavendish grade IV patients showed a mean decrease of 45° (range, 40°-60°) of SHABD.

Conclusion: Shoulder elevation is also impaired by GH joint contractures.

Sprengel's deformity, first described by Eulenberg in 1836, involves congenital scapular and periscapular dysplasia.[5] A scapula that is smaller and higher than normal and rotated inward, accompanied by taught periscapular tissues, results in limited shoulder girdle movement and an aesthetic defect.[2,4,8] Several surgical techniques have been proposed to improve function and physical appearance in these patients.[2,4,8]

A traditional axiom, believed to be first claimed by Fairbank in 1913, is that the shoulder elevation deficit observed in these patients is due to scapulothoracic joint stiffness.[6] Since then, hundreds of published papers have agreed with this claim, no studies having analyzed the role of the glenohumeral (GH) joint in shoulder motion restriction. It might be of interest in order to optimize treatment outcomes.

The purposes of this study were to (a) evaluate passive GH joint motion and (b) analyze its contribution to shoulder motion limitation in children with a Sprengel's deformity.

Methods

A prospective evaluation of 23 consecutive patients (8 girls and 15 boys, 15 right and 8 left) with a Sprengel's deformity, seen between April 2017 and April 2019, was performed. The following data were obtained by the same surgeon (FS): demographics, associated conditions, Cavendish grade of deformity severity, bilateral active global shoulder elevation, and multidirectional bilateral passive GH range of motion.[2] Passive GH measurements were performed with a goniometer and followed the methods described by Hodgson for children with neonatal brachial plexus

* Corresponding author: Francisco Soldado, MD, PhD, Pediatric Hand Surgery and Microsurgery Unit, Barcelona University Childrens Hospital HM Nens, Carrer del Consell de Cent, 437, 08009 Barcelona, Spain.
E-mail address: drsoldado@cirugiamanoinfantil.com (F. Soldado).

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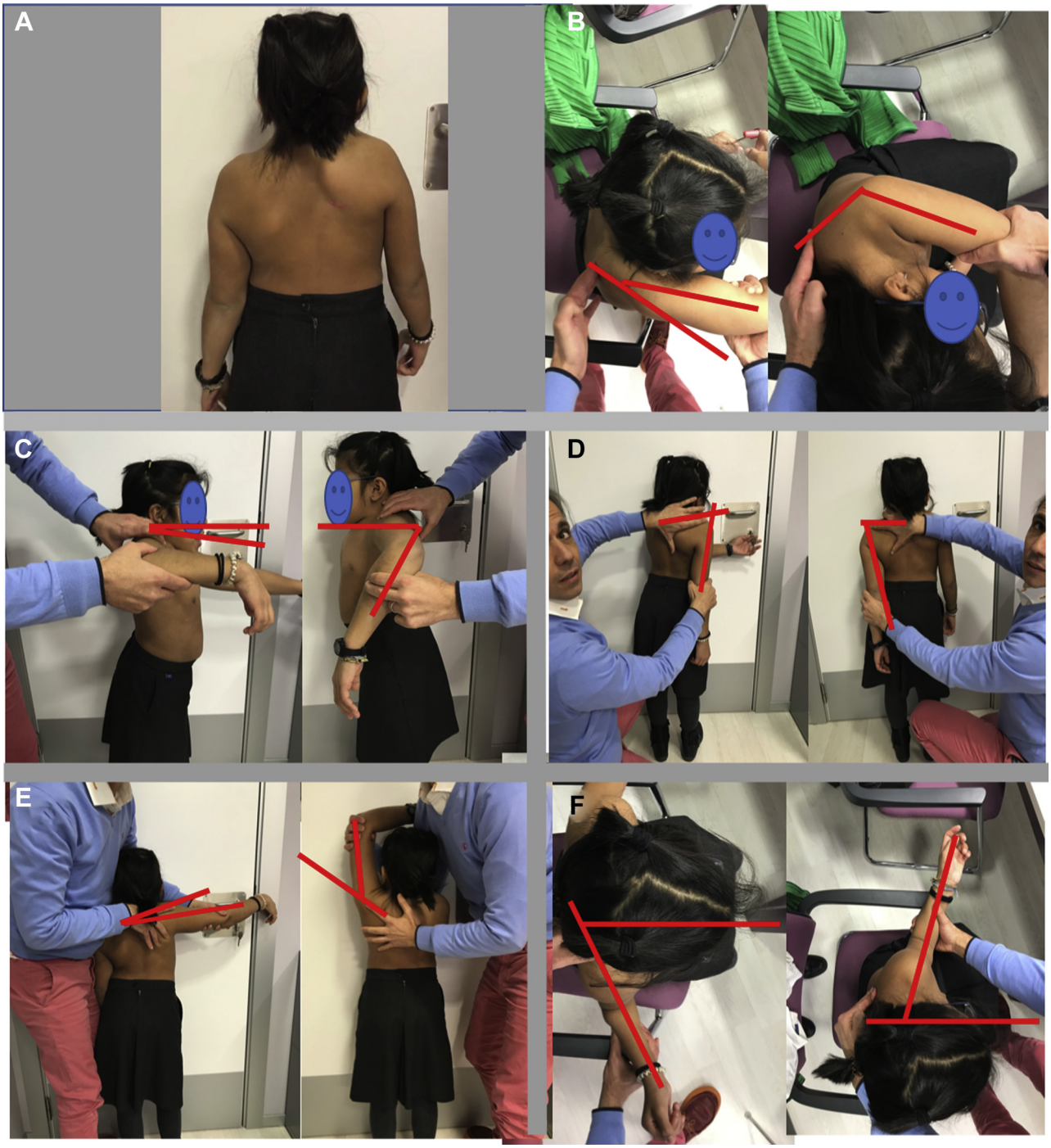

Figure 1 Passive range of glenohumeral motion following the Hodgson et al technique[9] in a 9-year-old child with a right Cavendish grade IV Sprengel deformity associated with Klippel-Feil syndrome (**A**). (**B,C**) Decreased right GH internal rotation in abduction and GH cross-body adduction, sign of posterior GH contracture. (**D**) Symmetric spinohumeral adduction angle showing no superior GH contracture. (**E**) Severe decreased right spinohumeral abduction angle, sign of inferior GH contracture. (**F**) Symmetric passive external rotation in shoulder adduction showing no anterior GH contracture. *GH*, glenohumeral.

injuries, including (Fig. 1) measuring GH internal rotation in abduction and GH cross-body adduction (CBADD) to assess for the degree of posterior GH contracture; spinohumeral abduction angle (SHABD) to assess the degree of inferior GH contracture; and spinohumeral adduction angle to assess the degree of superior GH contracture.[9] In addition, passive external rotation in shoulder adduction and abduction were measured to assess the degree of anterior GH contracture.[11] Patients provided written informed consent before participation in accor- dance with the Declaration of Helsinki guiding biomedical research involving human subjects.

Statistical methods

Data were summarized as means plus standard deviations for continuous variables and as absolute numbers with percentages for categorical variables. The Wilks-Shapiro test was used to determine the normality of continuous variables. For inferential comparisons, because patients' involved limb was being compared against their uninvolved limb, paired *t* tests were used. For correlation analyses, Pearson's correlation coefficients were calculated when both variables were continuous (age or degrees of active shoulder elevation)

and Spearman's correlation coefficients when either one of the paired variables was ordinal (Cavendish grade of deformity). An a priori decision was made to consider statistically significant absolute [r] values <0.30 as weakly correlated, from 0.30 to 0.69 as moderately correlated, and ≥0.70 as strongly correlated. To account for multiple comparisons, a Bonferroni-adjusted 2-tailed P value ≤.001 was set as the criterion for statistical significance. All analyses were performed using SPSS version 25 (IBM, Armonk, NY, USA).

Results

The mean patient age was 8.1 years (range, 1.4-16.7 years), and the Cavendish grade distribution was as follows: 13.4% grade 1 (n = 4), 52.2% grade 2 (n = 12), 13.4% grade 3 (n = 3), and 21.7% grade 4 (n = 5). All Cavendish type-4 cases were associated with Klippel-Feil syndrome.

There was a statistically significant decrease in the mean degrees of active global shoulder elevation on the involved vs. uninvolved side: 117.4 (range, 80-160) vs. 176.1 (range, 170-180), respectively; $P < .001$.

There were statistically significant decreases in the following involved vs. uninvolved mean passive GH measurements, all at $P < .001$: SHABD, 14.6° (range, −10° to 40°) and 41.5° (range, 30° to 50°); CBADD, 43° (range, 0° to 80°) and 71.3° (range, 60° to 100°); and internal rotation in abduction, 17.8° (range, 0° to 60°) and 49.4° (range, 10° to 90°), respectively. There were no statistically significant decreases in the following involved vs. uninvolved mean passive GH measurements: spinohumeral adduction angle, 19.1° (range, −10° to 40°) and 25.2° (range, 10° to 50°; $P = .069$); external rotation in shoulder adduction, 73.5° (range, 60° to 90°) and 78.5° (range, 60° to 90°; $P = .021$); and external rotation in shoulder abduction, 86.7° (range, 70° to 90°) and 90.6° (range, 90 ° to 110°), respectively ($P = .168$).

Strong inverse correlations were observed between the Cavendish grade (severity of the deformity) and both the extent of global shoulder elevation (r, −0.784) and SHABD (r, −0.669) (both $P < .001$). Cavendish grade was not correlated with any other passive GH movements. A strong inverse correlation also was evident between global shoulder elevation and SHABD (r, −0.677; $P < .001$). Global shoulder elevation was not correlated with other passive GH movements.

Cavendish grade IV patients exhibited a mean decrease of 91° (range, 80° to 100°) of shoulder elevation and 45° (range, 40° to 60°) of SHABD (Fig. 1).

None of the active or passive range of motion correlated with age.

Discussion

In our series of 23 patients with a Sprengel's deformity, GH joints on the involved side exhibited contractures both inferiorly and posteriorly. The more severe the deformity and decrease in shoulder elevation, the more severe was the inferior GH contracture. Thus, contrary to traditional beliefs,[6] the reduction in shoulder elevation was not just caused by scapulothoracic stiffness.

Glenohumeral stiffness in these patients might be congenital, due to the periscapular malformation itself, or might be acquired secondary to periscapular motion restriction. In our series, that there was no correlation between GH contracture severity and patient age suggests a congenital origin.

Glenohumeral stiffness might be due to muscular, capsular, or articular anomalies. Mears[12] has claimed that the long head of the triceps contributes to GH adduction restriction; thus, lengthening of this muscle head was added to the scapular osteotomy (Mears procedure). Cho et al,[3] using 3-dimensional computed tomography scans, identified no anomalies in the glenoid version in these children. Ogden et al[13] reported that the acromial end of the clavicle had a greater curvature on the involved side but did not appear to limit shoulder elevation.

Pathogenic considerations merit further study and are beyond the scope of the current report. Stiffness might be congenital, as suggested by the absence of correlation between age and GH range of motion measurements, or acquired due to an abnormal miotendinous or capsular growth in the absence of the physiological stretching derived form a normal scapular girdle motion. Glenohumeral stiffness in Sprengel's deformity patients is usually mild and might not have any clinical significance in most cases. However, marked inferior GH contracture was present in our 5 Cavendish grade 4 patients, all associated with Klippel-Feil syndrome, generating a mean loss of 45° of GH abduction. Thus, the worse reported results of corrective surgery in more complex cases might not be due only to scapulothoracic stiffness but also to GH anomalies that might be improved by adding surgical procedures addressing the GH joint.[1,4,10]

Posterior GH contracture, evidenced by decreases in CBADD and passive internal rotation in abduction, was also observed in our study.[15] This was not correlated with the severity of Sprengel's deformity, however. Although midline functional impairment was not measured in our patients, no patient complained of midline function problems.[7] Additional studies might analyze the clinical significance of posterior GH contractures in these children.

Our study had certain limitations apart from the limited number of patients. Among them is that, although we used clinical measurements of passive GH motion validated for congenital brachial plexus injuries,[9] due to the complexity of the shoulder girdle, laboratory motion analysis of both scapulothoracic and GH joints would be of benefit to confirm our results.[14]

Conclusion

Not only scapulothoracic limitations but also GH joint contractures contribute to functional deficits in patients with a Sprengel's deformity. The pathogenesis and potential treatment of these contractures merit further study.

Disclaimer

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