Correction in Malrotation of the Scapula and Muscle Transfer for the Management of Severe Sprengel's Deformity: Static and Dynamic Evaluation Using Three-Dimensional Computed Tomography

Background: The clinical results of surgical procedures for severe Sprengel's deformity have been uncertain. To obtain improved elevation, we consider that it is necessary to realign the lateral border of the scapula for upward rotation. The purposes of the current study were to evaluate the clinical results and range of motion of the scapula after such realignment.

Methods: Seven cases of Sprengel's deformity of Cavendish grade 3 or 4 were treated surgically and then clinically evaluated and examined using three-dimensional computed tomography (3D CT). (Two boys and five girls aged 50.9 ± 15.4 months, mean ± standard deviation at the time of operation.) The mean follow-up was 53.1 months (range, 12 to 92 months). After the omovertebral bone and the superomedial side of the scapula were removed, the levator scapulae and rhomboids were reattached to wrap around the scapula at maximum upward rotation to assist in maintaining this position. Cavendish and Rigault grades were used for evaluation of postoperative appearance. The superior displacement and rotation of the scapula were measured on the trunk posterior view using 3D CT. The relationship between improvement in the range of motion and radiological change were analyzed statistically.

Results: The postoperative flexion (97.9° ± 12.9° to 160° ± 11.5°) and abduction (99.3° ± 13.0° to 161.4° ± 15.7°) were significantly improved compared with the mean preoperative values ($P < .0001$). 3D CT revealed that in all patients the malrotation of the scapula was improved postoperatively. The current study shows that successful realignment of the scapula led to these improved clinical results.

Conclusions: Our procedure has advantages not only for recovery of the range of motion but also for reducing the characteristic lump in the web of the neck. However, our procedure has an inherent limitation related to asymmetrical shoulder level and width. 3D CT may be useful for preoperative planning and postoperative evaluation.

Level of Evidence: Level IV case series.

Key Words: Sprengel's deformity, muscle transfer, 3D CT

INTRODUCTION

Congenital elevation of the scapula, referred to as Sprengel's deformity, is a complex anomaly of malpositioning and dysplasia of the scapula with muscle hypoplasia and atrophy. Sprengel's deformity causes cosmetic deformity and functional limitations.[1, 2] Previous treatments have focused on the improvement of appearance, with functional improvement a secondary objective.[3, 4] Cavendish[3] noted that the primary aim of treatment is to improve appearance and that improvement in function is uncertain. However, various procedures have been developed to improve functional outcomes.[5-10] Green's procedure involves placing the scapula in an inferiorly located pocket and transferring the muscular attachments of the trapezius, rhomboids major and minor, and levator scapulae inferiorly.[5-7] Woodward first described the use of muscle transfers, and several modifications have been reported.[6, 8-10] From a functional viewpoint, abduction of the shoulder with this procedure is limited because of scapulothoracic stiffness and an inferiorly rotated glenoid.[11] Previous studies have reported that the angle of upward rotation ranged from 29° to 49° at 120° of elevation in the scapular plane.[12-14] To obtain an improved elevation, we consider that it is necessary not only to translate the scapula inferiorly but also to realign the lateral border of the scapula by upward rotation. Our strategy for improving functional problems associated with Sprengel's deformity is to manage the dynamic motion by upward rotation of the scapula. In our operative technique, we focus on achieving the space for upward rotation of the lateral border of the scapula, and on balancing muscle tension at the maximum shoulder elevation. The purposes of the current study were to evaluate the clinical results and, using three-dimensional computed tomography (3D CT), the motion of the scapula after its realignment and to assess the relationship between elevation of the shoulder and upward rotation of the lateral border of the scapula.

MATERIALS AND METHODS

Seven children were surgically treated in our hospital by one senior author between 2002 and 2010. The patients included two boys and five girls aged 50.9 ± 15.4 months (mean ± standard deviation) at the time of operation (Table 1). The mean follow-up was 53.1 months (range, 12 to 92 months). All patients had associated anomalies, including Klippel–Feil syndrome in three patients, Waardenburg syndrome in one patient, and Russell–Silver syndrome in one patient. Two had the complication of spina bifida and hemivertebra of the thoracic spine.

Surgical Procedure

For the current study, the surgical indication was moderate to severe restriction of the shoulder range of motion (less than 120°). The operation was performed with the patient under general anesthesia in a prone or a half lateral decubitus position. The affected scapula was exposed through a transverse incision on the scapular spine along Langer's lines. The trapezius, rhomboids and levator scapulae were detached from the scapular spine subperiosteally and the omovertebral bone or cord-like structure was excised extraperiosteally (Fig. 1a). **We always take care to protect the accessory nerve, which descends on the trapeziums in line with the medial border of the scapula. Several authors[3, 15] have recommended morselization of the clavicle in the Woodward operation. In our procedure, we focus on the realignment of the rotation of the scapula. The realignment of the scapula will reduce the risk of the brachial plexus palsy.** After exposure of the medial border of the scapula with detachment of the supraspinatus, infraspinatus and subscapularis muscles, the superomedial side of the scapula was removed to make space for maximum upward rotation of the scapula. The scapula was then realigned at a position of maximum elevation. In this study, the long head of the triceps and a portion of the origin of the teres minor were not resected from the scapula. The levator scapulae and rhomboids were reattached to the medial border of the scapula with drill holes and nonabsorbable sutures at the maximum upward rotation of the scapula to help to maintain this position (Fig. 1b). The supraspinatus, infraspinatus and subscapularis muscles were repaired to wrap around the scapula. The excised trapezius was sutured to its insertion at the scapular spine. The origin of the middle part of the trapezius was transferred inferiorly by two vertebral levels, except in two patients with hypoplasia of the inferior trapezius muscle (#6, 7), where the trapezius muscle transfer was not performed. A sling was applied during the first postoperative week, and pendulum exercises were started the day after surgery. Active movements of the shoulder were allowed 2–3 days postoperatively.

Clinical Evaluation and 3D CT

Clinically, the improvement in the range of motion and in the Cavendish[3] (Table 2) and Rigault[16] grades (Table 3) were recorded at final examination. In radiographic assessment, the level of the superior angle of the scapula was measured on the trunk posterior view with reference to the thoracic spine. An omovertebral bone was found in four patients and a fibrous cord-like structure was found in one patient. In the first four patients, postoperative CT scans (Aplio, Toshiba Medical Systems Corp., Otawara, Japan) in both a resting potion and at maximum elevation were performed. In the last two patients, CT scans in both positions were performed both prior to surgery and postoperatively. The obtained 3D images were reconstructed with a slice thickness of 2 mm at increments of 2 mm. The superior displacement and rotation of the scapula were measured on the trunk posterior view using 3D CT according to a modification of a previously described method.[5] The superior displacement of the affected scapula was measured by the difference between the superior scapular level and the contralateral scapular level (Fig. 2). The scapular length was measured from the superior angle to the inferior angle. The angle between the line of the scapular spine and the thoracic spine was measured as the rotation angle of the scapula (Fig. 2).

Statistical Analyses

Data for continuous variables are reported in the text as means ± standard deviations. The mean changes between preoperative and postoperative flexion and abduction were compared using paired *t*-tests. A Pearson correlation coefficient was calculated and a scatter plot was drawn to show the association between elevation of the humerus and rotation angle of the scapula. All reported *P* values are two-sided and those under 0.05 were considered to be significant. Data analysis was performed using JMP ver. 9 (SAS Institute, Cary, NC, USA).

RESULTS

The mean preoperative flexion improved significantly postoperatively (97.9° ± 12.9° to 160° ± 11.5°), as did abduction (99.3° ± 13.0° to 161.4° ± 15.7°) ($P < 0.0001$) (Table 4). The cosmetic appearance improved postoperatively by one or two levels on both the Cavendish and Rigault classifications (Fig. 3). Visible deformity as a lump in the web of the neck was classified as Cavendish grade 2. In our series, three of seven patients had an improved appearance of the lump after surgery. The scar cured without keloid except in the patient with Waardenburg Syndrome (Table 1). **It is hard to state that there was no case any nerve injury at all, because almost all the patients have hypoplastic muscles including trapezius and rotator cuff. However, the range of motion in all patients improved after the surgery. Moreover, no patients have complained about difficulty in daily activities after the surgery. Therefore we conclude there was no severe nerve**

injury. Although the straps of a backpack sometimes fall off the patient's shoulder because of the short shoulder girdle, no patients reported any impairment in performing daily activities. No recurrence of the omovertebral bone and no regeneration of the resected part of the scapula have been encountered in any patient at the final examination.

In a 3D CT evaluation, the mean superior displacement of the affected scapula decreased postoperatively (Table 4). The mean rotation angle of the scapula at the resting position increased from 81.6° ± 18.7° preoperatively to 98.0° ± 11.8° postoperatively. Although it could be measured in only two patients, the rotation angle of the scapula at the elevated position increased from 106.5° ± 26.2° preoperatively to 137.2° ± 13.9° postoperatively. The rotation angle of the scapula improved postoperatively in all patients. The mean rotation angle ratios of the scapula at rest and elevation, which postoperatively were 0.89 and 0.92, respectively, indicated a more symmetrical upward rotation of the lateral border of the scapula postoperatively compared with the preoperative findings (Fig. 4). The relationship between elevation of the humerus and upward rotation angle of the scapula was 0.76 (P = 0.026, Fig. 5).

DISCUSSION

Our procedure, resection of the omovertebral bone and multiple muscle transfer, has enabled an improved upward rotation angle of the scapula. The 3D analyses demonstrated that the rotation angle of the scapula improved postoperatively in all patients. The present study showed that realignment of the rotation of the scapula has excellent or satisfactory clinical results. The mean postoperative flexion and abduction improved to 160.0° and 161.4°, respectively. The relationship between elevation of the humerus and the rotation angle of the scapula was 0.76. These results indicate that successful scapular realignment led to better clinical results.

In our operative procedure, muscle balance is a key factor for the static and dynamic realignment of the scapula. We focused on the correction of scapular malalignment for functional improvement, especially in patients with a severely limited range of motion. Previous studies have noted muscle malformations involving the trapezius, the rhomboids, and the levator scapulae.[3, 10] Transfer of these malformed muscles may lead to insufficient improvement of the rotation of the scapula. One of the advantages of our procedure is the ability to manage the balance of transferred muscle and to adjust the appropriate scapular resection during the operation, even though some muscles are hypoplastic. The direct change of the point of application on the trapezius, levator scapulae, and rhomboids provides appropriate tensioning of the transferred muscles. The superior effect of transfer of the inferior trapezius muscle at the origin has already been recognized in the Woodward procedure.[8] In our series, transfer of the trapezius muscle at the origin was performed in five shoulders. Thus, multiple muscle transfer is the basis of our procedure.

We found significant improvement not only in function, but also in cosmetic appearance after operation. All patients improved at least one level on the Cavendish and Rigault scales. Our study showed that the scapular realignment gave an unremarkable superomedial part of the scapula and a cosmetic improvement. However, we recognize that our procedure has an inherent difficulty in achieving symmetrical shoulders. The shoulder width and the scapular shape remain asymmetrical even after surgery. To solve these problems, additional treatment is required. Moreover, realignment of the upward rotation of the lateral border of the scapula risks having a postoperative glenoid level higher than the preoperative level. Our procedure is limited in its ability to recover the depression of the scapular position. In the case of severe scoliosis, achieving elevation with upward rotation does not always lead to sufficient improvement in abduction. Although the scapular angle and the scapular angle ratio at elevation in patient #7 improved to 124° and 0.79, respectively, abduction only improved to 130°.

We compared the scapular motion at rest and in an elevated position by measuring the scapular spine angle using 3D CT. Cho et al reported morphometric analysis of Sprengel's deformity using 3D CT.[17] Visualization of scapular motion using 3D CT provides a better understanding of the effectiveness of each operative procedure. In all patients, the scapular angle improved postoperatively. The scapular angle ratio was 0.89 at rest and 0.92 at elevation, indicating that static and dynamic realignment of rotation of the scapula after the operation was successful and that the transferred muscles were effective for scapular rotation. The Cavendish grade is based on the cosmetic condition and the Rigault scale is based on radiographs. However, some patients showed a good range of motion despite unsatisfactory cosmetic or radiographic presentation. To obtain improved elevation, we considered that it was necessary to realign the upward rotation of the lateral border of the scapula. Therefore, we needed to evaluate the scapular angle correctly. This 3D CT study showed a strong relationship between the elevation of the humerus and the rotation angle of the scapula.

There were several limitations in our study. First, it included only a small number of patients with heterogeneous background variables, such as age, anomalies, and follow-up period. These factors could affect our results. The age of the patients must be taken into account. Only children over six years of age are suitable for scapular transplantation.[18] Second, we did not evaluate the 3D motion of the scapula, such as retraction, even using the 3D model. A final consideration is the radiation exposure at a young age. In this series, each patient was exposed to 2–3 mSv in each 3D CT. Therefore, 3D CT would not be appropriate for regular follow-up evaluation because of this high radiation exposure.

In conclusion, resection of the omovertebral bone and multiple muscle transfer has allowed improvement of the upward rotation angle of the scapula. 3D CT evaluation showed successful realignment of rotation after surgery. Our procedure has advantages not only for functional recovery but also for reducing the characteristic lump in the web of the neck. However, shoulder level and width remained asymmetrical. Although 3D CT evaluation has some limitations, it could be useful for preoperative planning and postoperative evaluation.

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Figure Legends

FIGURE 1. Surgical procedure. (a) Resection of the medial border and transfer of the trapezius, rhomboids, and levator scapulae for realignment of rotation of the scapula. OMB: omovertebral bone; Tra: trapezius; Rho: rhomboids; LS: levator scapulae. (b) Postoperative scapular position. The levator scapulae and rhomboids were reattached at the maximum upward rotation of the scapula.

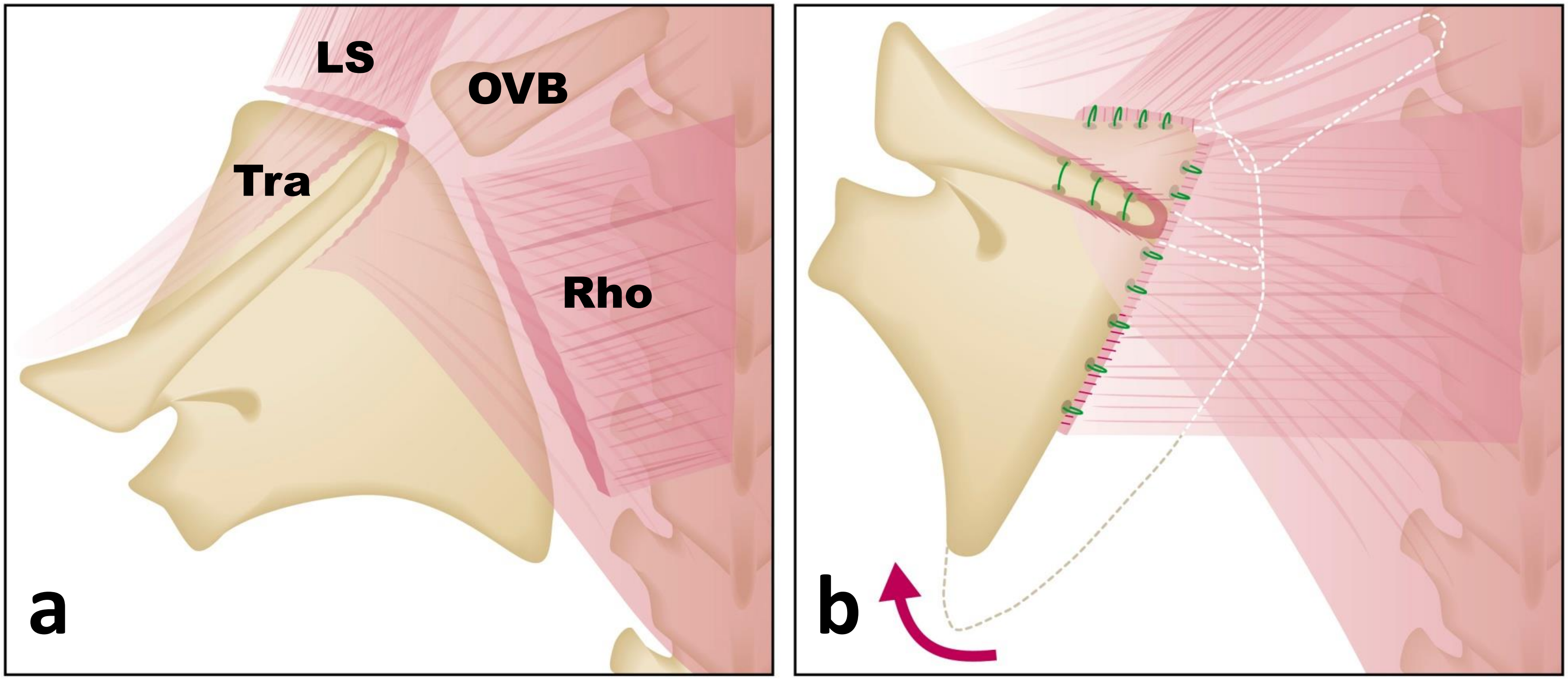
FIGURE 2. Measurement of the superior displacement and rotation of the scapula assessed by 3D CT. I: The distance between the line from the superomedial angle of the scapula perpendicular to the vertebral axis line in affected and unaffected shoulders. II: the scapular length in the unaffected shoulder. Superior displacement = I/II. III: the rotation angle of the scapula is between the line of the scapular spine and the vertebral axis. Rotation angle ratio of the scapula = affected/unaffected.

FIGURE 3. Case 2 at the age of three years with preoperative Cavendish grade 4. (a) The preoperative appearance. (b) Note the preoperative limitation of abduction on the affected side. (c) The postoperative deformity was classified as Cavendish grade 2 after four years operation. Surgical scarring was unremarkable. (d) Note the improvement of shoulder function and cosmetic appearance.

FIGURE 4. Case 6 at the age of seven years with preoperative Cavendish grade 3. (a) 3D CT of the preoperative rotational deformity at rest. Note the superior displacement on the affected side compared with the unaffected side. (b) Preoperative at elevation. Note the lesser rotation angle of the scapula on the affected side compared with the unaffected side. (c) Postoperative at rest. (d) Postoperative at elevation. Note the superior migration, and that the rotation angle of the scapula has improved postoperatively.

FIGURE 5. A Pearson correlation coefficient was calculated and a scatter plot was drawn to show the association between elevation of the humerus and the rotation angle of the scapula.

The correlation between elevation of the humerus and upward rotation angle of the scapula was 0.76 ($P = 0.026$).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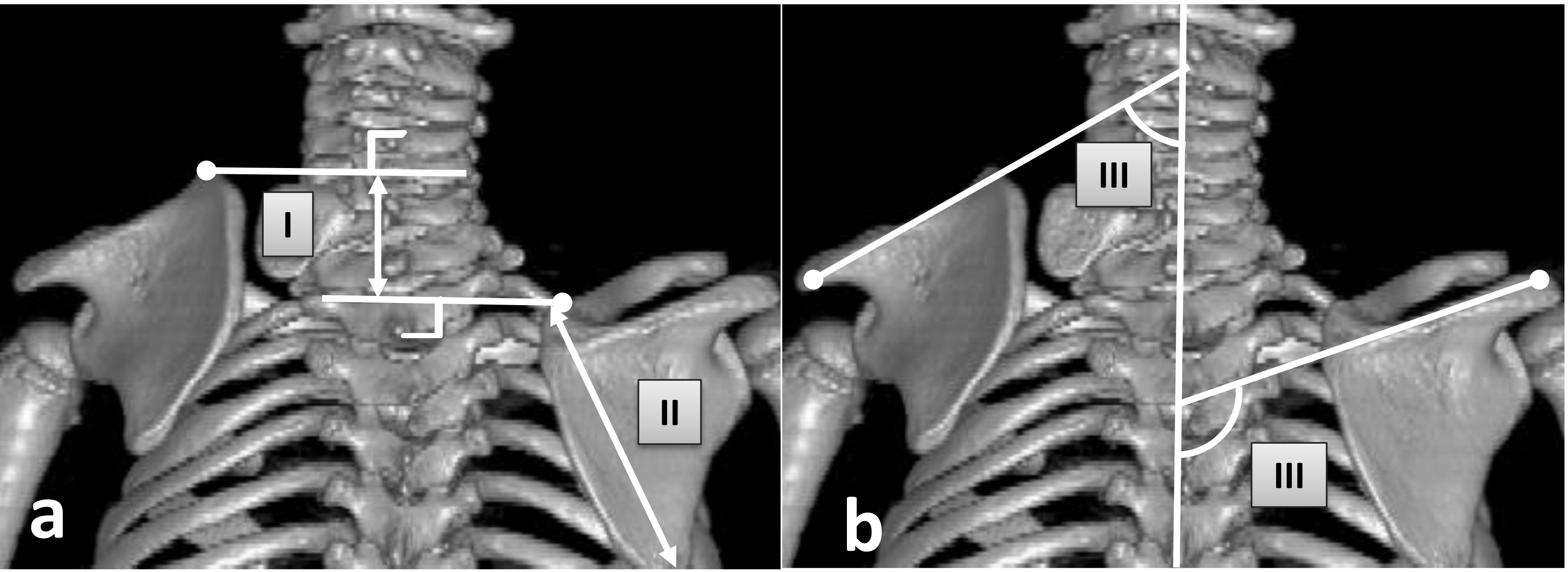
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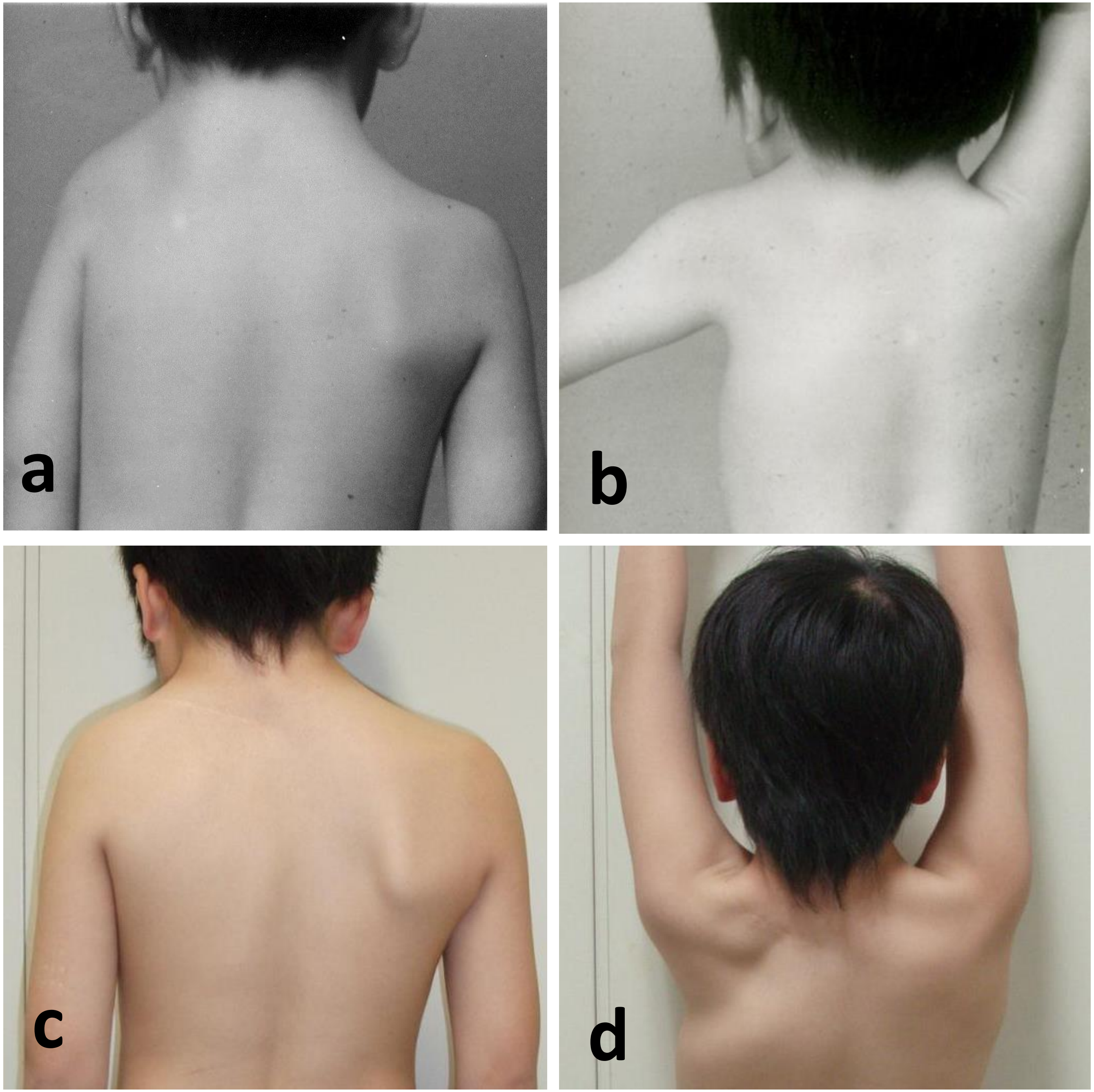
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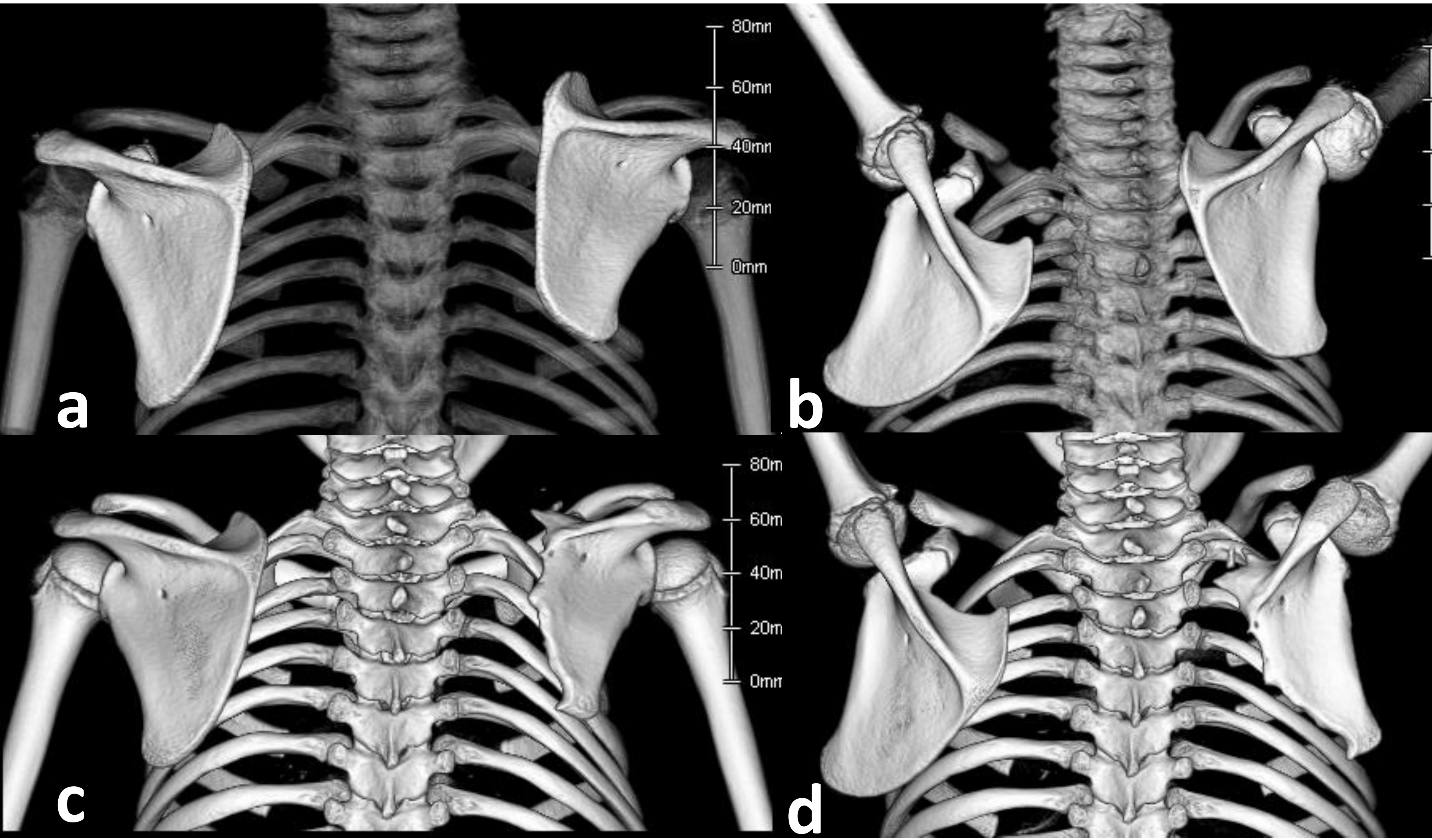

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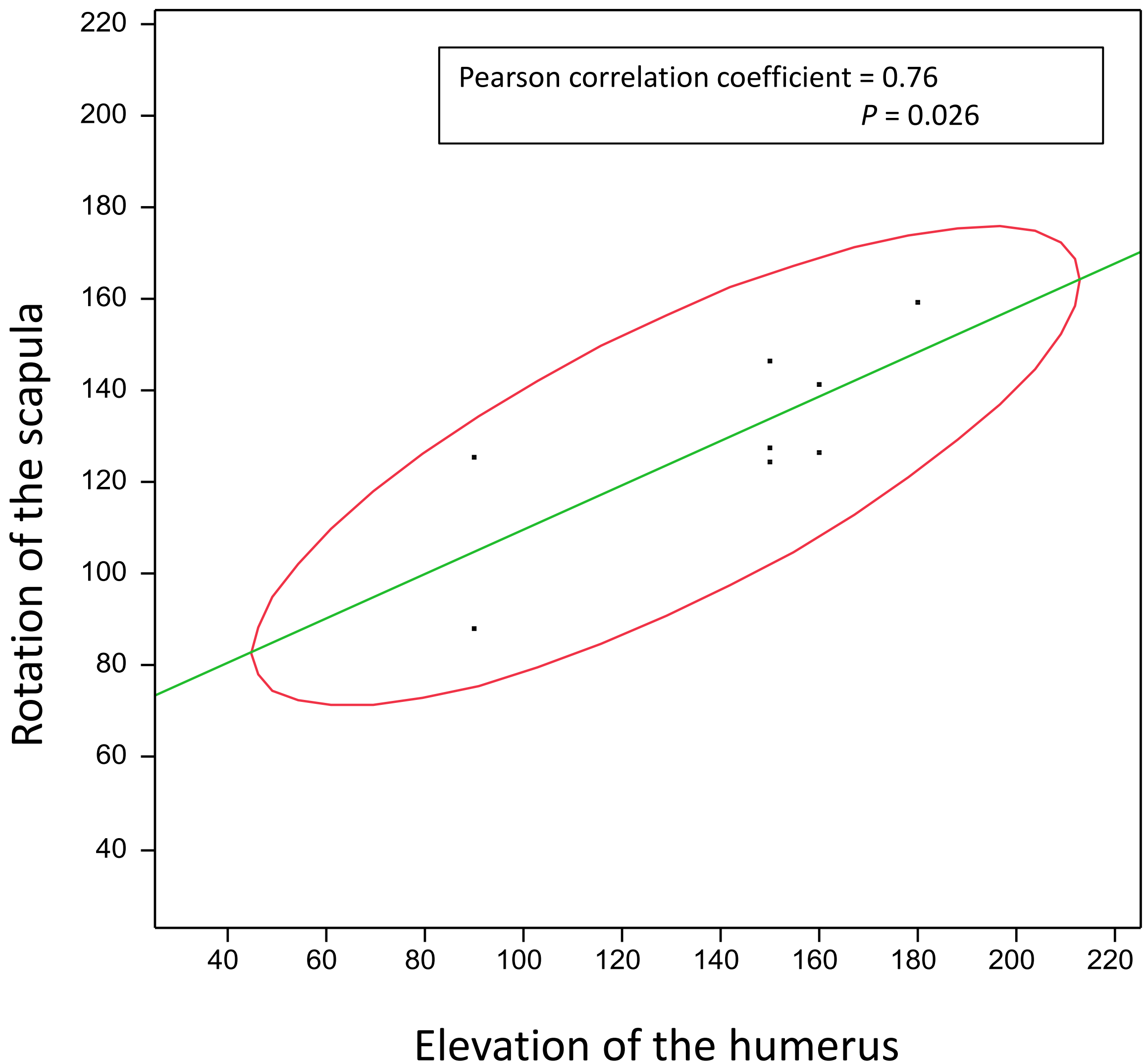

Pearson correlation coefficient = 0.76
P = 0.026
Rotation of the scapula
Elevation of the humerus
220
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