

## Case Report

# Three-year follow-up after Woodward procedure in children with Sprengel's deformity

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### ABSTRACT

Sprengel deformity is a rare congenital disorder characterized by a congenital high scapula caused by obstruction of the development of the scapula, which accompanies bone, muscle and cartilage during pregnancy. This scapular deformity can be associated with other congenital malformations such as rib abnormalities, scoliosis, and Klippel-Feil syndrome. This is a follow-up of 3-year-old children with Sprengel deformity, cavendish grade 3 who were treated with the Woodward procedure. Authors conclude that the Woodward procedure is an effective procedure for obtaining good functional and cosmetic results.

**Keywords:** High scapula, Sprengel's deformity, Undescended scapula, Woodward procedure

### INTRODUCTION

Sprengel's deformity is a rare congenital disorder, first described by Eulenberg in 1863, and in 1891 by Sprengel.<sup>1,2</sup> In this deformity, the scapula is elevated, hypoplastic, and usually somewhat adducted.<sup>3</sup> Attention is drawn to the rotation of the scapula that makes the glenoid facing down and the superior angle forms a distinctive lump in the neck tissue.<sup>4</sup>

This congenital disorder is caused by disruption of the development of the scapula in the normal caudal migration of the scapula at the beginning of fetal development so that the scapula elevated and hypoplastic.<sup>5,6</sup> This scapular deformity can be associated with other congenital abnormalities such as rib disorders, scoliosis, Klippel-Feil syndrome and in some cases spina bifida.<sup>7</sup>

In 20-25% of patients, omo-vertebral bone is present, either consisting of bone, cartilage, or fibrous tissue,

between the superior angle of the scapula and the apophysis of the spinous process of the cervical vertebrae.<sup>4,8</sup>

Patients usually have asymmetrical shoulders and limited shoulder abduction. Abduction is limited to the shoulder joint caused by limited movement of the scapula due to the presence of the omo-vertebral bone or cartilage structure between medial border the scapula and the vertebral spinous process in the lower cervical spine.<sup>7</sup>

Authors report a case of a 3-year follow-up of a 3-year-old boy with a Sprengel deformity who was treated with the Woodward procedure.

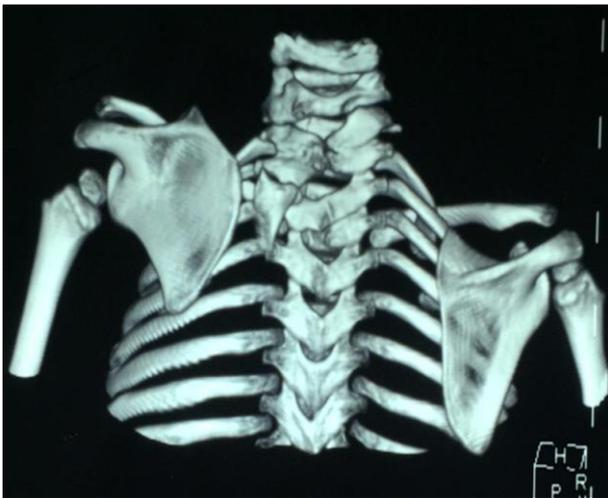
### CASE REPORT

A 3-year-old boy was admitted to our hospital because of the abnormal appearance of the left shoulder and limited shoulder abduction (Figure 1).



**Figure 1: Left shoulder before surgery, abduction 90.**

The patient cannot abduct the left shoulder more than 90 degrees. This disorder is considered by parents since the patient was born.



**Figure 2: CT Scan showed asymmetric shape of the scapula with omo-vertebral bone.**

From the radiography and CT scan there is an asymmetrical shape of the left scapula, the left scapula is smaller and located higher than the right scapula, and also there is an omo-vertebral bone located between the medial border of the left scapula and the spinous process (Figure 2).

Authors diagnosed this patient with a left Sprengel's deformity, with a grade 3 cavendish classification, based on elevated of the shoulder joint that was more than 2 cm, and the deformity was easily visible.

Authors performed operative treatment for this patient, using a Woodward procedure, an incision was made from the spinous process of C1 to the spinous process of Th 9, the greater and lesser rhomboid muscles were detached, and the omo-vertebral bone was completely excised. The trapezius muscle was detached from its origin on the spinous processes of the vertebrae, the scapula was

displaced distally, and the origins of the rhomboid and trapezius muscles were reattached to the spinous processes more distally.

After surgery authors put velpeau shoulder bandage for three weeks to provide comfort to patient, and to prevent excessive movement during the healing process. Patient recovered well without complication and velpeau shoulder bandage were removed after three weeks and continued to various exercises and shoulder physiotherapy.



**Figure 3: Two-month after surgery, abduction 130.**

Two months after surgery (Figure 3) there was an improvement of the shoulder cosmetic and function, the shoulder asymmetry almost level, and the Cavendish classification increased to grade 1, which is the shoulder joint are level, and invincible deformity when the patient was dressed.



**Figure 4: Three-year follow-up, abduction 150.**

The abduction function of the shoulder increased, with the acquisition of abduction 40 degrees, the patient can abduct a total of 130 degrees, before surgery the patient can only abduct up to 90 degrees. After 3 years of follow-

up (Figure 4), there is a further increase in shoulder abduction of 20 degrees, therefore abduction is 150 degrees, with acceptable scars.

## DISCUSSION

Various procedures have been described to improve the appearance and function of the shoulder of patients with Sprengel deformities.

However, the preferred method has been the Woodward procedure, since it does not require a postoperative spica cast and much lower risk of bleeding and brachial plexus injury.<sup>9</sup> The Woodward procedure has been praised in having the ability to provide better both cosmetic and functional results.<sup>10</sup>

Woodward procedure is basically a gentle procedure which carries a low risk of brachial plexus palsy and profuse bleeding. It is based on scapular transplantation to a lower level by moving the spinal origins of the rhomboid and trapezius muscles caudally.<sup>7,11</sup>

The modifications in the Woodward procedure including additional excision of the medial border of the scapula to prevent postoperative apparent winging of the scapula by decreasing its horizontal width.<sup>12</sup>

Surgical treatment is indicated in mild and moderate cases (grades 2 and 3) because it provides improvement in both function and appearance.<sup>7</sup> Whereas in other study surgical intervention is best indicated for moderate and severe condition with significant cosmetic deformity (grades 3 and 4) and significant limitation of shoulder abduction.<sup>13</sup>

The contemporary results of Woodward procedure showed a positive correlation of functional and cosmetic outcomes through short and long terms, with the procedure and more improvement occurs in cases of younger age at the time of surgery.<sup>14</sup>

Previous studies furthermore reported the best time for surgery to be between 3 and 8 years of age, whereas Jeannopoulos advocates surgery for children between 2 and 5 years of age.<sup>7,15</sup>

At a younger age, the procedure is likely to be too difficult. After 8 years of age, no substantial improvement might be expected due to the combination of growth, loss of biological plasticity of soft tissues or fibrosis, loss of function due to fixed structures.<sup>3,4,9,10</sup>

This method produced marked improvement in appearance, as assessed on the cavendish scale. The improvement of abduction ranged from 0 to 60 degrees.<sup>16</sup> Earlier studies on the results of the Woodward procedure reported a mean functional improvement in abduction of 32 to 59 degrees and few complications.<sup>3,4,8-10,12</sup>

In our case, authors corrected about 60 degrees, without complication. The scar heals well, and parents are quite happy with the results of treatment.

## CONCLUSION

The Woodward procedure is effective in achieving functional and cosmetic improvements in children with Sprengel's deformity.

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