Adult presentation of congenital muscular torticollis is rare. We report 12 patients with this condition who underwent a modified Ferkel's release comprising a bipolar release of sternocleidomastoid with Z-lengthening. They had a mean age of 24 years (17 to 31) and were followed up for a minimum of two years. Post-operatively a cervical collar was applied for three weeks with intermittent supervised active assisted exercises for six weeks.

Outcome was assessed using a modified Lee score and a Cheng and Tang score. The mean pre-operative rotational deficit was 8.25° (0° to 15°) and mean lateral flexion deficit was 20.42° (15° to 30°), which improved after treatment to a mean of 1.67° (0° to 5°) and 7.0° (4° to 14°) after treatment, respectively. According to the modified Lee scoring system, six patients had excellent results, two had good results and four had fair results, and using the Cheng and Tang score, eight patients had excellent results and four had good results.

Surgical management of adult patients with neglected congenital muscular torticollis using a modified Ferkel's bipolar release gives excellent results. The range of neck movement and head tilt improved in all 12 patients and cosmesis improved in 11, despite the long-standing nature of the deformity.

Con genital muscular torticollis is common. Of congenital musculoskeletal abnormalities only hip dysplasia and talipes equinovarus are more common.1,2 Its frequency in the newborn ranges from 0.3% to 2%.3 It involves contracture and shortening of the sternocleidomastoid muscle, causing the head to tilt towards the affected side and the chin to the opposite side.1,4 The suggested causes include trauma during birth, ischaemia, intrauterine malposition, constitutional and growth arrest causes, compartment syndrome, hereditary predisposition and neurogenesis.1,2,4 The child commonly presents with swelling and deformity in the first week of life.3 Untreated, the sequelae include facial asymmetry, tight bands, ophthalmic abnormalities and altered neck posture.5 The main reason for consultation is cosmetic deformity of the head and face. Treatment includes non-operative means such as stretching exercises, physiotherapy, and botulinum toxin injections.1,6 with surgical options comprising subcutaneous and open tenotomies,7 resection and Z-lengthening of the sternocleidomastoid muscle.1,8,9 In resistant cases, the extent of sternocleidomastoid tightness determines the choice of unipolar or bipolar lengthening, Z-lengthening or radical resection.1,10,11 Akazawa et al11 also reported partial resection of sternocleidomastoid and reported acceptable results and less recurrence. Recently, Lee et al12 also reported good results in 20 patients, but observed loss of strength of sternocleidomastoid and alteration in the contour as limitations of this method. Z-lengthening of the affected muscle can give good results, but it is difficult to allow for subsequent growth in children,11 and it may lead to tethering and recurrent deformity.11 Other studies have shown that Z-lengthening is not essential in older children.2,11 Distal unipolar lengthening is sufficient for most cases, but secondary or bipolar lengthening may be needed, especially with a recurrent problem.1,4 Muscle release and lengthening procedures can maintain muscle strength and provide better cosmesis.3 A high rate of recurrence after surgical release has been described by some authors.7,8,11,14 Bipolar release involves release of both inferior and superior attachments and can be a good option for patients over six years of age and in neglected cases.5,12,13,15

Parameters such as residual head tilt, scar formation, craniofacial asymmetry and age at the time of surgery play an important role in the outcome after surgery.2,4,8 Good or excellent results have been reported following surgery in between 40% and 88% of cases.4,11,12,16 The age of the patient affects...
both the natural course and the outcome, and the ideal age for surgery is still debated. Ling felt the ideal age to be between one to four years, and suggested that after five years of age, the risk of complications increased. With a delay in presentation, factors such as craniofacial asymmetry may become irreversible, but good results from surgical intervention in older children have been reported. Long-standing congenital muscular torticollis and presentation in adults is rare, as most deformities are managed in childhood. Few cases of correction in adulthood are reported in the literature. Earlier reports concluded that surgery for adult torticollis is hazardous, although later studies have reported good results in terms of range of movement of the neck and cosmetic correction.

This study was undertaken to assess the clinical and functional outcome in a series of adult patients with neglected congenital muscular torticollis treated surgically.

Patients and Methods

Between 2005 and 2008, 14 adults with neglected congenital muscular torticollis presented to the senior author (SP). Two with ophthalmic pathology (congenital nystagmus) were excluded, leaving 12 patients (nine men and three women) with a mean age of 24.0 years (17 to 31) in the study (Table I). The right side was involved in seven and the left in five. Anteroposterior and lateral cervical radiographs of these patients were normal, with no degenerative changes or vertebral abnormalities. The passive range of movement of rotational and lateral flexion was measured clinically with a goniometer by the senior author (SP) and compared with that of the opposite side in order to calculate the deficit.

Operative technique. All patients underwent a modified Ferkel’s procedure with bipolar release of sternocleidomastoid by the senior author.

Under general anaesthesia with the patient supine the neck is turned to the opposite side. A small bolster is placed under the ipsilateral shoulder. Two incisions are used. A small retro-auricular incision is made over the mastoid and the proximal head of sternocleidomastoid is identified as a broad, shiny insertion on the mastoid process. The tendon is freed from bone and allowed to slide distally. A second incision is made over the medial end of the clavicle. After dividing the platysma, the sternal and clavicular heads are exposed. Fibrous bands in the fascia are divided. The clavicular head is more muscular and is completely separated from bone. Z-lengthening up to the mid-portion of the tendon of the sternal head of sternocleidomastoid is performed and the neck is gently manipulated to stretch the sternocleidomastoid. The tendon ends are approximated with 3-0 monofilament absorbable sutures in the stretched position to preserve the cosmetic V-shape of the neck. Subcuticular closure is done in layers to reduce the likelihood of scar formation. Postoperatively a hard collar is worn for three weeks. Gradual supervised stretching and lateral flexion exercises, out of the collar, are started as comfort permits and continued for six weeks.

The final results were compiled and statistically evaluated by two authors (SP, AKS), including the modified Lee scoring system which assessed both function and cosmesis, scored from 0 to 3. Function was assessed by movement of the neck. Cosmesis was assessed by head tilt, scarring and lateral band formation. The modified Cheng and Tang Score was also used. This involves measurement of the rotational and lateral flexion deficit in degrees measured with a goniometer comparing the normal side to the affected side, and assessment of craniofacial asymmetry (none, mild, moderate, severe), scar (none, mild, moderate, severe), lateral band (none, lateral, clavicular, sternal), subjective assessment (cosmetic and functional) and head tilt in degrees measured clinically with a goniometer on a frontal photograph of the patient. All headings were scored from 0 to 3 according to severity, and a total score of 17 to 21 was considered excellent, from 12 to 16 good, 7 to 11 fair, and less than 7 was poor.
Statistical analysis. The changes in the mean ranges of movement following surgery were compared for statistical significance using Wilcoxon’s signed ranks test. A p-value < 0.05 was considered significant.

Results
The mean follow-up was 2.96 years (two to four). The pre-operative mean rotational deficit was 8.25° (0° to 15°) and mean lateral flexion deficit was 20.42° (15° to 30°) which was reduced to a mean of 1.67° (0° to 5°) and 7.0° (4° to 14°), respectively (Table I). These improvements were statistically significant (both p < 0.001). According to the modified Lee scoring system, six patients had an excellent result, two had a good result and four had a fair result (Table II). The appearance of the scar was good in 11 patients, with no adhesions or hypertrophy; one graded the cosmetic improvement as poor. No patient experienced a complication or required further surgery for residual bands. Four patients (cases 3, 5, 8 and 9) had a persistent rotational deficit of 5° and three patients (3, 5 and 7) had a persistent lateral flexion deficit of ≥ 10°. According to the Cheng and Tang score, eight patients had an excellent and four had a good result.

Discussion
The primary goals of surgery for patients with congenital muscular torticollis are improved movement of the neck and an improvement in head tilt. Cosmetic improvement is also an important consideration. According to these criteria we were successful in managing all our patients, despite the long-standing nature of the deformity, except one with a poor cosmetic outcome.

Neglected congenital muscular torticollis is rare in adults. Socioeconomic reasons and restricted provision of healthcare may be a cause for the deformity remaining untreated into adulthood. In India, girls are generally brought for correction of deformity just prior to their wedding, as was the case with all three women in our series, but this was also the reason for one of the men (case 4) being brought for treatment. In these late presentations craniofacial asymmetry may be irreversible owing to loss of remodelling potential. Nevertheless, correction of the soft-tissue deformity can give a good cosmetic result.

The timing of surgery for this condition remains controversial. Early release has been advised by some to avoid craniofacial asymmetry and to reduce the length and expense of treatment. Surgical correction after one year of age has been advised because of the need for compliance...
with post-operative rehabilitation.\(^1\) Contraindications to early treatment include concerns in relation to infection, wound breakdown and haematoma formation.\(^2\) Ling\(^3\) suggested that the optimal timing for correction was one to four years of age. Lawrence and Azizkhan\(^4\) found better results in children operated on after one year, but before two years. Chen and Ko\(^5\) and Minamitani, Inoue and Okuno\(^6\) concluded that surgery for this condition in children over six years of age was safe but others concluded that surgery after ten years of age was less rewarding.\(^7,19,26\) This contrasts with the experience of Sudesh et al,\(^8\) who reported good results in children older than ten. Other authors have reported that surgical correction up to 12 years of age is safe.\(^5,27,28\) Shim and Jang\(^9\) suggested that the most important factor in determining the ideal time for surgery was compliance with rehabilitation, rather than the age of the patient. Only a few reports are available of the outcome in patients with neglected congenital torticollis.\(^7,10,13,18,29,30\) These studies include patients to a maximum age of 36 years, with all obtaining benefit.

Omid-Kashani et al,\(^2\) in a prospective study of 14 neglected cases of congenital torticollis, concluded that bipolar sternocleidomastoid division, even in patients with irreversible facial asymmetry, improves quality of life by improving neck movement and head tilt. Employing two systems of scoring, all our patients had an excellent, good or fair outcome. The reduction in the mean deficit in the range of movement indicated good clinical outcome. Only one patient (case 3) graded his cosmetic improvement as poor, despite a good range of movement.

Bipolar release has been suggested as the operation of choice for patients with neglected congenital torticollis.\(^4,5,8,13,31\) The only controversial issue is Z-lengthening of the sternocleidomastoid. If a bipolar release is performed without Z-lengthening, loss of the normal contour of the neck may affect the cosmetic appearance.\(^16\) It has been claimed that with meticulous repair of the platysma and soft tissues no loss of normal contour occurs.\(^2,13\) In our series we performed bipolar release with Z-lengthening in all cases, with all patients reporting improved movement and head tilt and good cosmetic results in all but one. Hollier et al\(^20\) suggested that in older patients there was a requirement for a more extensive release, including trapezius, but resection of a part of the sternocleidomastoid has been proposed in longstanding cases.\(^2\) We did not need these additional measures to achieve correction in our patients.

Different post-operative protocols have been employed in these patients, including casts, braces, traction, early exercises and even halo vests.\(^4,19,24\) Immobilisation in an overcorrected position has been claimed to give better results;\(^17\) others have advocated the long-term use of braces to prevent recurrence,\(^7\) or intermittent cervical traction in the early post-operative period followed by a cervical collar and exercises.\(^19\) We followed a simple programme of supervised post-operative bracing and exercise, with good results.

Our study has limitations. The sample size is small, but neglected congenital muscular torticollis presenting in adulthood is unusual. The rotational and lateral flexion deficit was assessed by a single assessor (SP), and the consistency of the measurements was not investigated. We did not specifically measure facial asymmetry. However, our patients reported improved self-perception of facial asymmetry and most had a good cosmetic result.

In conclusion, surgical management of adults with neglected congenital muscular torticollis using a bipolar release of sternocleidomastoid and Z-lengthening gives excellent clinical and functional results.

### Supplementary material

A table detailing the complete results according to the Cheng and Tang score and pre- and three-year post-operative clinical photographs of patient no. 11 are available with the electronic version of this paper on our website at www.jbjs.org.uk

The authors wish to thank the Indian Orthopaedic Research Group for technical help in reviewing the literature and revision of the manuscript. We would also like to thank N. Jhangiani for English proofreading of the final manuscript.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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