CASE REPORT

Bryan artificial cervical disc arthroplasty in a patient with Klippel-Feil syndrome

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Technological advances have made more options available for surgical intervention in spinal disorders. From spinal fusion to artificial disc implantation, these advancements have brought great benefits, allowing preservation of spinal motion and flexibility after intervertebral discectomy. Yet the use of artificial discs as a treatment for congenital spinal disorders has been documented in only a handful of publications. We report a case where a Bryan artificial cervical disc arthroplasty was used to maintain and preserve the mobility and function of the cervical motion segments adjacent to fused vertebral lesions in a 33-year-old woman with Klippel-Feil syndrome who presented with chronic neck pain and signs of early myelopathy. The rationales for using the Bryan disc prosthesis system in patients with Klippel-Feil syndrome and its advantages over conventional surgical interventions are discussed.

Introduction

The treatment of cervical spine disorders is undergoing a paradigm change from favouring fusion to motion preservation. Anterior cervical discectomy with fusion (ACDF) was a well-accepted procedure in the past but long-term studies have shown a 25% prevalence of adjacent segment symptoms within 10 years. This is due to increased biochemical stresses and accelerated degeneration of the neighbouring spinal motion segments.[1] In recent years, much effort has been made to investigate the use of the artificial disc systems as an alternative to fusion. It is well-documented that the immediate and short-term results of artificial disc arthroplasty are equivalent to those of ACDF.[2] Disc arthroplasty has the additional advantages of being able to maintain cervical motion and reduce the incidence of adjacent segment degeneration. Since the implantation of the first Bryan cervical disc (Medtronics Sofamor Danek, Memphis, Egypt) in January 2000, more than 5000 patients have been treated with the Bryan disc.[3] It has been used in a wide spectrum of cervical spine disorders from trauma to spondylotic myelopathy. Sekhon[4] has reported the use of the Bryan disc in the management of adjacent segment degeneration associated with previous fusion surgery and surgery at the cervicothoracic junction. Nevertheless, no cases of artificial cervical arthroplasty in patients with congenital spinal diseases like the Klippel-Feil syndrome have been reported. It is widely believed that arthroplasty may be difficult to perform owing to the anatomical anomalies and shape of the vertebrae interfering with instrumentation and positioning during the procedure. We report the first case of use of the Bryan disc arthroplasty in a patient with Klippel-Feil syndrome who presented with symptoms of cervical myelopathy.

Case report

The patient, a 33-year-old woman, presented with a 2-year history of slowly progressive neck pain and headache. The neck pain was localised to the posterior midline with bilateral radiation to her shoulders and occiput up to the vertex, disturbing her sleep and work. Oral analgesics had been tried without much improvement. She had bilateral hand numbness and clumsiness that made her unable to work as a waitress. Physical examination revealed a mildly decreased range of movement in her neck, bilateral positive Hoffman's signs, and early myelopathic signs (hyperreflexia) in her lower limbs. There was bilateral diminished touch sensation on her fingertips. A cervical spine X-ray showed multiple abnormalities: there was partial incorporation of the C1 vertebra to the base of skull; C2/3 vertebral bodies showed incomplete segmentation and fusion of the posterior elements; C6/7 vertebral bodies were also fused with a segmentation abnormality on the right side involving the posterior elements. On flexion/extension views there was an increase in the atlanto-axial distance of more than 6 mm, suggestive of C1/2 subluxation. Magnetic resonance imaging (MRI) confirmed the same bony anomaly with C1 occipitilisation, incomplete C2/3 segmentation, and complete C6/7 fusion (block vertebrae). There were

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採用Bryan人工頸椎間盤置換手術治療先天性頸椎缺少或融合

技術不斷發展，使治療頸椎病的手術種類愈來愈多，從脊柱融合術到人工椎間盤植入，這些新發展的技術造福病人，使他們在接受椎間盤切除手術後，脊柱仍能保留活動能力並屈伸自如，但人工椎間盤用於治療先天性頸椎病的文獻紀錄並不多見。本病例報告記述一名33歲女患者，因先天性頸椎缺少或融合而長期頸痛，並出現早期脊髓病，她接受Bryan人工頸椎間盤置換手術，使鄰近出現融合椎體病變位置的頸椎運動節段能保留原有的功能和機動性。報告亦探討了採用Bryan人工頸椎間盤系統治療先天性頸椎缺少或融合的理據，以及該術比傳統手術優勝之處。

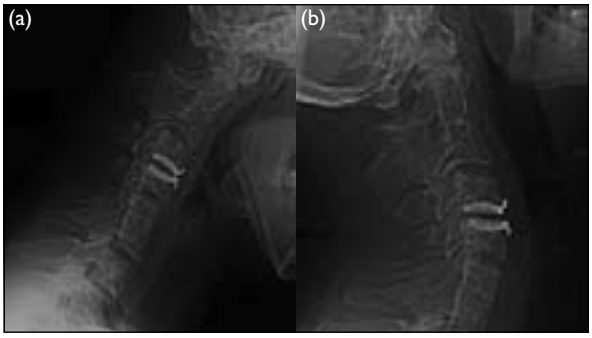

FIG 2. Lateral cervical spine X-rays before (a) and after (b) flexion with the Bryan disc arthroplasty at the C4/5 level demonstrating preservation of motion after operation
Note the adjacent vertebral abnormalities: fused C6/7, incomplete C2/3 segmentation and C1 occipitalisation

FIG 1. The Bryan artificial disc prosthesis

posterior disc bulges at the C3/4 and C4/5 levels. The thecal sac was indented with effacement of the anterior and posterior cerebral spinal fluid spaces at C4/5. There was cord flattening at this level but no accompanying abnormal cord signal changes.

Her symptoms persisted despite physiotherapy and rest. Surgical intervention was indicated because of the significant clinical and radiological myelopathic signs and the risk of further deterioration. There were two issues to tackle. The first was her C2 nerve root pain secondary to C1-2 subluxation and the second was her C4/5 myelopathy. In order to prevent cord damage during positioning for the posterior fusion for C0-2, decompression of the C4/5 disc protrusion was considered first. In view of the significant loss of motion that would occur with a traditional ACDF of C4/5 combined with posterior fusion of the upper cervical levels, an anterior decompression with an artificial disc arthroplasty at the C4/5 level was adopted.

Surgical technique

A preoperative computed tomographic (CT) scan of the spine was done to assess the anatomy of the cervical vertebrae and thus confirm the feasibility of an arthroplasty. With the use of the Bryan Cervical Disc Prosthesis System (Medtronics, Inc, Memphis, Egypt) [Fig 1], an anterior cervical discectomy at C4/5 was performed, and a 14-mm artificial disc prosthesis was then placed at this level under radiological guidance. The implantation technique followed the established procedural protocol.[3] The total operating time was 2 hours and 30 minutes and there were no surgical complications. No cervical collar was placed.

Postoperative flexion/extension X-rays (Fig 2) and MRI of the cervical spine demonstrated maintenance of motion at the level of instrumentation with adequate decompression of C4/5 and satisfactory placement of the prosthesis.

A posterior C0-C2 fusion with an iliac crest bone graft was performed 6 weeks afterwards for the C1/2 instability. The surgical procedure was well-tolerated, with postoperative imaging showing satisfactory alignment and preservation of cervical spine mobility. The patient showed improved symptoms with less hand numbness and was able to return to work after 2 weeks' rest.

Discussion

Klippel-Feil syndrome occurs in a heterogeneous group of patients unified only by the presence of a congenital defect in the formation or segmentation of the cervical spine. Numerous associated abnormalities including congenital scoliosis, deafness, Sprengel's deformity, and genitourinary and cardiovascular

abnormalities, may be present.

Treatment regimens depend on the severity of symptomatic segmental instability or neurological compromise, varying from modification of activities to extensive spinal surgery.[5] Occipitocervical arthrodesis is accomplished most commonly via a posterior approach. In atlanto-axial and sub-axial arthrodeses, wires are commonly used to achieve atlanto-axial fusion. When posterior elements are deficient, lateral mass, transpedicle, or transarticular screw-plate fixations have been performed but these have left significant limitations of movement afterwards. Anterior decompressions with arthrodeses have been carried out for affixed sagittal deformities. The concept of disc replacement is not new. Stainless-steel (metal-on-metal) ball-in-socket devices have been implanted in cervical motion segments adjacent to Klippel-Feil lesions, and these have been found to preserve motion for more than a decade following surgery.[6] With its newer metal-polymer-metal design, the Bryan disc prosthesis aims to provide the full range of coupled movements within a sub-axial cervical motion segment, as well as offer a load-dampening effect similar to that of a natural disc. The Bryan disc is a composite-type artificial disc designed with a low friction, wear resistant, elastic nucleus with two anatomically shaped metal plates. A flexible membrane forms a sealed space and contains a lubricant to reduce friction and wear and tear. This implant allows a normal range of motion and comes in five sizes. Technically the procedure used for the Bryan disc arthroplasty is similar to that of other disc-replacement operations, except that it requires milling of the end-plates, thereby generating more bone dust. This problem can be overcome by using more irrigation during the operation and anti-inflammatory drugs postoperatively. Heterotrophic ossification has been reportedly associated with Bryan disc arthroplasty.[7]

We considered an arthroplasty with the Bryan artificial disc the most suitable choice for our patient because of several factors. Firstly, our patient is young and preservation of the motion in her remaining cervical spine segments is important for maintaining neck function and quality of life. Her multi-level disease means the segments without a segmentation abnormality bear the main shearing forces during flexion and extension of the neck. That accounts for the most significant degenerative spondylotic changes seen at the C3 to C5 levels. An ACDF would have restricted the range of movement in her entire cervical spine. The Bryan disc is effective according to data from the prospective European human clinical trial (January 2000), which showed preservation of a mean (standard deviation) of 9° (5°) of motion at the 2-year follow-up evaluation.[8] Secondly, cervical arthroplasty is appropriate in our patient because at the C4/5 level there was predominantly anterior rather than posterior compression of the spinal cord and there was no significant cervical stenosis behind the vertebral body which would have made a cervical corpectomy a more appropriate choice. Thirdly, adjacent-segment degeneration associated with a traditional ACDF should be avoided, as this would aggravate the degeneration in the spondylotic spine and necessitate another operation to treat these transitional levels.[9] A CT of the cervical spine was needed before the operation to study the bony anatomy and the feasibility of accommodating the Bryan disc in view of the highly variable features of this congenital anomaly. The main issue with the Bryan disc is its durability and long-term effects on adjacent motion segments, which remain as yet undetermined although it has been tested under laboratory conditions to 45 human equivalent years of neck movement with only little wear noted. Furthermore, the maintenance of motion should theoretically impose less biochemical stress on the adjacent levels compared with fusion.

Conclusion

The categories of patients receiving artificial disc arthroplasties have been expanding with improvements in techniques and the emergence of more clinical outcome data. We report the first case where an artificial disc arthroplasty has been performed in a patient with the Klippel-Feil syndrome, where a congenital defect occurs in the cervical spine segmentation. Decompression of the most spondylotic level was achieved, and motion was maintained. The clinical and radiological outcomes were satisfactory, but long-term follow-up is required to evaluate the functional preservation achieved by this method. For patients with congenital spinal disease, careful evaluation of the correlation between the anatomical anomaly and functional compromise is important when considering the feasibility of arthroplasty in this group.

References

1. Bridwell KH, Anderson PA, Boden SD, Vaccaro AR, Zigler JE. What's new in spine surgery? J Bone Joint Surg Am 2004;86-A:1587-96.
2. Goffin J, Van Calenbergh F, van Loon J, et al. Intermediate follow-up after treatment of degenerative disc disease with the Bryan Cervical Disc Prosthesis: single-level and bi-level. Spine 2003;28:2673-8.
3. Wang MY, Leung CH, Casey AT. Cervical arthroplasty with the Bryan disc. Neurosurgery 2005;56(1 Suppl):58S-65S.

4. Sekhon L. Cervicothoracic junction arthroplasty after previous fusion surgery for adjacent segment degeneration: case report. Neurosurgery 2005;56(1 Suppl):E205.
5. Tracy MR, Dormans JP, Kusumi K. Klippel-Feil syndrome: clinical features and current understanding of etiology. Clin Orthop Relat Res 2004;424:183-90.
6. Boden SD, Balderston RA, Heller JG, Hanley EN Jr, Zigler JE. An AOA critical issue. Disc replacements: this time will we really cure low-back and neck pain? J Bone Joint Surg Am 2004;86-A:411-22.
7. Leung C, Casey AT, Goffin J, et al. Clinical significance of heterotopic ossification in cervical disc replacement: a prospective multicenter clinical trial. Neurosurgery 2005;57:759-63.
8. Bryan VE Jr. Cervical motion segment replacement. Eur Spine J 2002;11(Suppl 2):92S-97S.
9. Hilibrand AS, Carlson GD, Palumbo MA, Jones PK, Bohlman HH. Radiculopathy and myelopathy at segments adjacent to the site of a previous anterior cervical arthrodesis. J Bone Joint Surg Am 1999;81:519-28.