

Surgical correction of rigid cervicothoracic deformity in a transgender patient: case report

Bilal B. Butt, Paul Gagnet, Joshua Piche, Rakesh Patel, Ilyas S. Aleem

Department of Orthopaedic Surgery, University of Michigan, Ann Arbor, MI, USA

Correspondence to: Ilyas S. Aleem, MD, MS, FRCSC. Department of Orthopedic Surgery, University of Michigan, 1500 East Medical Center Drive, 2912 Taubman Center, SPC 5328, Ann Arbor, MI 48109, USA. Email: ialeem@med.umich.edu.

Abstract: A number of spinal pathologies result in fusion of the spine, including ankylosing spondylitis, diffuse idiopathic skeletal hyperostosis (DISH), as well as severe degenerative arthropathies. This fusion of spinal elements may result in spinal deformity affecting any region of the spine. Cervicothoracic deformity resulting in chin on chest deformity is poorly tolerated due to inability to maintain a horizontal gaze. Surgical treatment options for this condition are complex and require extensive discussion between the patient and surgical team. Here we present a case report of a 26-year-old transgender female (male to female) patient with severe chin on chest deformity and a unique pattern of spinal fusion involving only the posterior elements. She underwent C2–T8 posterior spinal fusion with thoracic pedicle subtraction osteotomy and multiple cervical facet osteotomies with good functional result. She did have severe dysphagia and required feeding tube for several weeks but did very well by 1 year postoperatively. While posterior elements of the spine are normally affected first in spondyloarthropathies such as ankylosing spondylitis, the lack of anterior spinal involvement is unique and could be attributed to hormonal therapy in this patient. This case describes a unique pattern of spondyloarthropathy and highlights the importance of having a multi-disciplinary team for the treatment of patients with complex spinal pathologies.

Keywords: Ankylosing spondylitis; spondyloarthropathy; cervical kyphosis; chin on chest deformity; pedicle subtraction osteotomy; transgender; case report

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Introduction

The cervical spine is an essential anatomic structure that protects neurologic elements and is fundamental for maintenance of horizontal gaze. Normal alignment of the cervical spine, particularly in the sagittal plane, is imperative for normal physiologic function and low muscle energy expenditure. Cervicothoracic kyphosis is a spinal condition which moves the center of gravity anteriorly, resulting in increased energy expenditure and decreased ability to maintain horizontal gaze. Furthermore, this may result in cervical myelopathy secondary to spinal cord stretching and alteration in microcirculation (1). Spondyloarthropathies, such as ankylosing spondylitis, can result in anomalous fusion of the spine and is a potential etiology for spinal deformity, resulting in significant patient morbidity and

impact on overall quality of life.

Due to inflammatory processes that first affect the facet joints, patients with ankylosing spondylitis begin to flex their spine to offload contact pressures. Bridging of syndesmophytes occur, eventually leading to ankylosis and the characteristic bamboo spine appearance on radiographs (2,3). This eventually leads to patients developing a fixed kyphotic deformity that is largely centered at the cervicothoracic and thoracolumbar junctions (4). In an attempt to compensate for the deformity, patients flex their knees and extend their hips to restore horizontal gaze, though this requires profound energy expenditure and cannot be sustained for prolonged periods of time (5). Several radiographic measures are used to measure and monitor the cervicothoracic deformity, including the Chin-

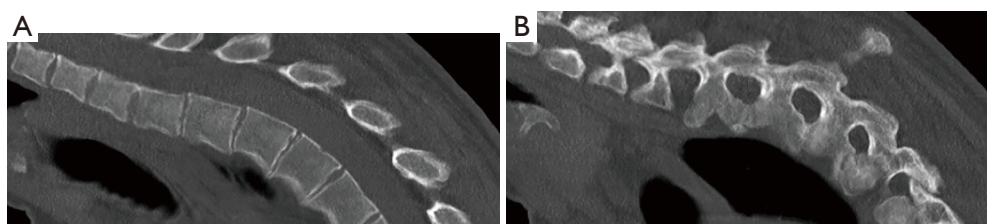


Figure 1 Select CT slices showing unfused anterior segments and fused posterior segments of the spine. (A) Sagittal CT scan of cervicothoracic junction with unfused anterior elements of the spine; (B) sagittal CT scan of cervicothoracic junction with fused posterior elements of spine.

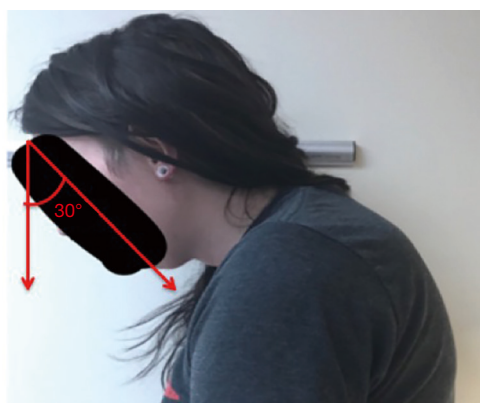


Figure 2 Clinical photo demonstrating CBVA of 30 degrees.

Brow vertical angle (CBVA), the C2–C7 sagittal vertical axis (SVA), and C2–C7 angle (6). We report a unique surgical case of spondyloarthropathy affecting the posterior spinal elements only, resulting in severe chin-on-chest deformity in a transgender patient. The authors present the case in accordance with the CARE reporting checklist (available at <http://dx.doi.org/10.21037/jss-20-584>).

Case presentation

The patient is a 26-year-old transgender female (male to female) with progressive kyphotic cervicothoracic deformity due to spinal fusion of posterior elements only (*Figure 1*). As a child, she underwent Nuss bar placement for pectus excavatum, and this was subsequently removed. She was found to be HLA-B27 positive and was being treated with adalimumab. For her gender transition, she was being treated with high dose estrogens and anti-androgens. She did undergo pre-operative bone health evaluation and was found to have a lumbar Z score of -0.1 . Clinically, she demonstrated a near chin on chest deformity with a rigid, fixed kyphotic deformity (*Figure 2*). She was very



Figure 3 Lateral standing radiograph of spine, with C7 plumb line, demonstrating negative sagittal balance.

unhappy with her inability to look straight ahead. Imaging demonstrated a C2–C7 SVA of 8.7 cm and a C2 to C7 angle of 13.7 degrees (*Figures 3,4*). After exhausting non-operative treatment, and after extensive discussions about

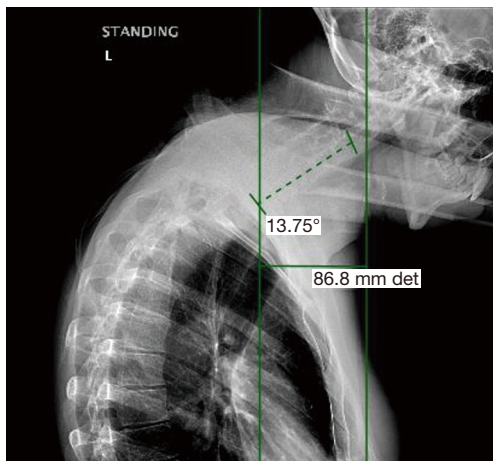


Figure 4 Lateral radiograph of cervical spine demonstrating C2–C7 angle of 13.7 degrees and a C2–C7 SVA of 8.7 cm.



Figure 5 Three-dimensional image, demonstrating T2 PSO with posterior spinal fusion from C2–T8. Note- resection of portion of ribs bilaterally. Staples present distally.

surgical correction and the associated risks with her, as well as her rheumatologist and family, surgical intervention was agreed upon. She underwent a C2–T8 posterior spinal fusion with a T3 pedicle subtraction osteotomy (PSO) and C3–7 cervical facet osteotomies. This location was chosen as the optimal osteotomy location as it was felt to be the apex of the deformity and thus provided the site for the largest correction of her deformity. In briefly reviewing the various types of osteotomies, we felt the PSO would provide the largest correction per level. In the PSO a closing wedge osteotomy of the vertebral body occurs with extension through the pedicle. This is held in place with dorsal spinal instrumentation. Uniquely, thoracic pedicle subtraction osteotomies, such as hers, often require partial rib resections

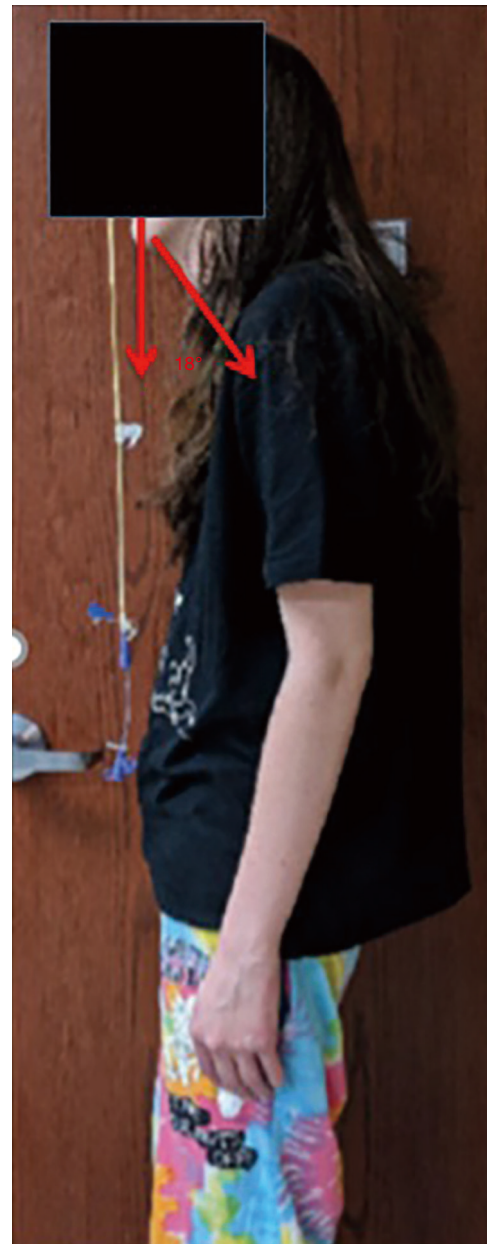


Figure 6 Post-operative lateral clinical image, demonstrating CBVA of 18 degrees. Note, Dobhoff tube in place to help with difficulty swallowing in acute post-operative period.

(Figure 5).

She had no intraoperative complications and was very happy with her postoperative outcome and restoration of horizontal gaze. However, she did require a Dobhoff tube for feeding due to severe dysphagia (Figure 6). Nutritionist was consulted and optimized her intake to ensure adequate

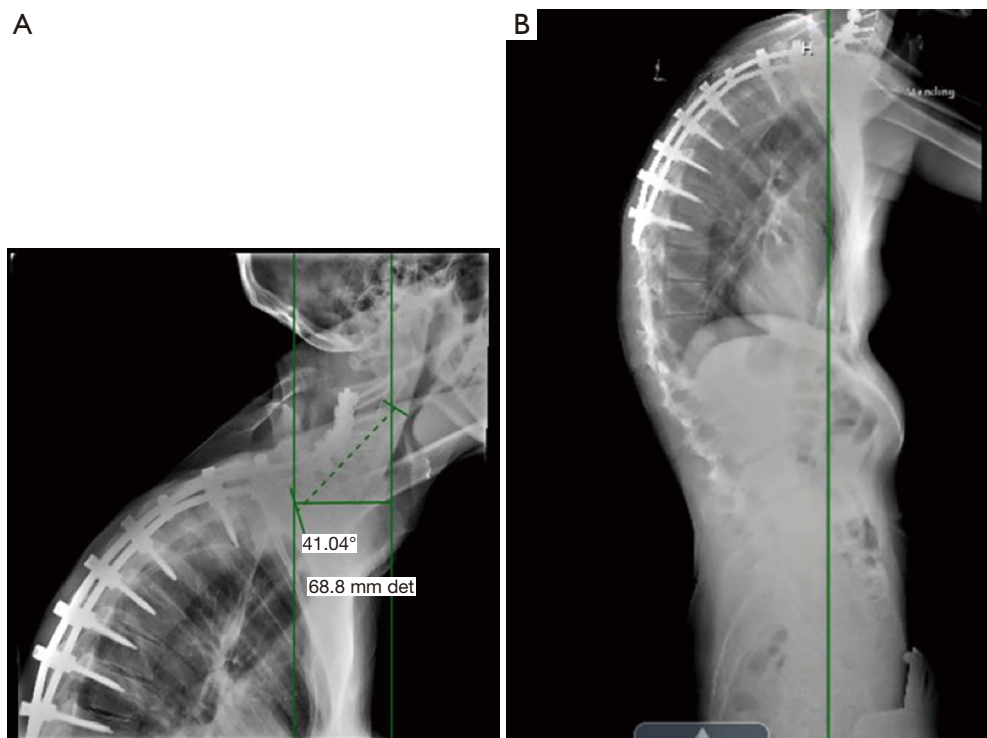


Figure 7 Postoperative cervical and whole spine radiographs. (A) Lateral cervical radiograph demonstrating C2–C7 SVA of 6.8 cm, and a C2–C7 angle of 41 degrees; (B) post-operative lateral radiograph demonstrating improvement in SVA. Pedicle screw fixation noted from C2–T8.

nutrients. By her 3-month post-operative visit, this was removed and she was able to tolerate oral intake well. By her 1-year post-operative visit, she was performing activities she enjoyed without restrictions, though she did report occasional muscular aches, and reported an improved quality of life. Postoperative 1-year imaging is shown in (Figure 7). There were substantial improvements in the C2–C7 SVA and C2–C7 angle, as well as her global alignment.

Consent was obtained from this patient for publication of this case report.

Discussion

Ankylosing spondylitis (AS) is a systemic inflammatory disease that predominately affects the axial skeleton leading to pain, deformity, and in some instances neurological deficits (2,7-9). Deformity of the spine can occur in all regions of the spine, though the kyphotic deformity of the cervical spine is poorly tolerated due to inability to maintain a horizontal gaze. AS is primarily seen in white males of non-Asian/African heritage (7,9). Symptoms begin to

develop in most patients when they are in their twenties. While there is no single known cause of the disease there, 90% of patients with AS are found to be positive for Human Leukocyte Antigen subtype B-27 (HLA-B27) (7,10). Attempts at understanding the role that androgens play in the pathogenesis of AS is not yet fully elucidated, but there is belief that men have more severe disease, and faster progression of the disease (11-13).

This case report is the first to report correction of complex deformity in a transgender patient. Many studies have demonstrated low bone mass in trans-women and recommend pre-operative bone health evaluations when patients have elevated risk factors. These risk factors include; initiation/termination of hormone therapy especially post gonadectomy, prior fragility fracture, and low levels of vitamin D (14-16). It is imperative to evaluate bone health in this patient population prior to deformity correction to identify any underlying metabolic abnormality that may affect bone density. Evidence regarding rates and risk factors of spinal fusion in patients on hormone replacement or therapy is lacking and a focus for future

research.

Goals for successful correction of cervical kyphosis are maintenance of comfortable horizontal gaze, decompression of neurological elements, and stable fusion of the head aligned with the pelvis. Once the location of the osteotomy is chosen, a standard dorsal approach to the spine is performed. Once the spine is exposed the authors prefer to utilize intra-operative navigation to place pedicle screws or lateral mass screws cranial and caudal to the osteotomy site. Hardware is not placed at the intended osteotomy site. A complete laminectomy is performed, along with removal of the superior articulating facet and inferior articulating facet. The exposed pedicle remains intact and is decancellated using a burr. From this decancellated pedicle the vertebral body is decancellated, and eventually the pedicle is amputated at the pedicle body junction. Using a Woodson or similar tool, a plane is established between the remaining body and the spinal cord. The cortex of the body is impacted into the decancellated portion of the vertebral body, creating a defect just anterior the spinal cord. Under direct visualization the defect in the body is closed, ensure no kinking of the cord. In the thoracic spine the ribs are partially removed, to allow for closure of the vertebral body defect. If they are not removed, they prevent closure of the osteotomy site. This closure is held in place with the instrumentation that was placed prior to the osteotomy.

A number of complications may occur following complex cervical deformity correction, including infection, new neurologic deficit, nonunion, or distal junctional kyphosis. One of the most common and most debilitating, however, is dysphagia. It is imperative to counsel patients about this preoperatively. Due to the severity of the kyphotic deformity in patients undergoing major cervical surgery, many already have baseline subclinical swallowing dysfunction (17). It is important to establish baseline level of function with evaluation by an otolaryngologist, especially if patients have any complaints of dysphagia preoperatively. Post-operatively many patients report clinically significant dysphagia, and there should be a low threshold for multi-disciplinary approach with speech language pathology and nutritional consult to assess safe swallowing and metabolic needs to aid in bone healing (18,19).

Conclusions

Cervicothoracic deformity is a complex pathology resulting in significant patient morbidity and decreased quality of life. Surgical correction aims to restore sagittal alignment but

has a high complication profile. Patients must be warned that complications are likely, though severity can vary from trouble swallowing to neurological compromise (20,21). Our patient presented a number of unique challenges, including a complex rigid cervicothoracic deformity in the context of posterior spinal element fusion only. While posterior elements of the spine are normally affected first in spondyloarthropathies such as ankylosing spondylitis, the lack of anterior spinal involvement is unique and could be attributed to hormonal therapy in this patient. Alternatively, this may be a unique pattern of spinal fusion not yet completely described in the literature.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist (available at <http://dx.doi.org/10.21037/jss-20-584>).

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at: <http://dx.doi.org/10.21037/jss-20-584>). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Consent was obtained from this patient for publication of this case report.

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