Case Report

Surgical management of Bertolotti’s syndrome in two adolescents and literature review

Christopher E. Louie¹, Jennifer Hong², David F. Bauer¹, ²

¹Geisel School of Medicine at Dartmouth, Hanover, ²Department of Neurosurgery, Dartmouth-Hitchcock Medical Center, Lebanon, New Hampshire, USA.

E-mail: Christopher E. Louie - christopher.e.louie.med@dartmouth.edu; Jennifer Hong - jennifer.hong@hitchcock.org

*David F. Bauer - david.f.bauer@hitchcock.org

INTRODUCTION

Bertolotti’s syndrome is defined as a congenital lumbosacral transitional vertebra (LSTV) that is responsible for disabling low back pain. It most commonly occurs at the L5 level, followed by the L6 level, and is characterized by various morphologic presentations [Table 1]. Several medical and surgical therapies are available to treat this syndrome: for example, physical therapy, corticosteroid injections (many risks/complications without documented long-term efficacy), laminectomy, spinal fusion, and removal of the pathologic bone segment. Here, we demonstrate the outcomes after surgical resection of LSTV in two pediatric patients and have reviewed the relevant literature.

CLINICAL PRESENTATION

Patient 1

A 14-year-old female with midline low back pain and the right hip/leg pain was treated for 9 months with physical therapy and nonsteroidal anti-inflammatory drugs (NSAIDs) without...
recovery. Her neurological examination was normal. Lumbar X-ray, computed tomography (CT), and magnetic resonance imaging (MRI) studies showed Bertolotti's syndrome, characterized by a right-sided partially sacralized L5 vertebra with pseudoarthrosis between L5 to S1 and ileum, with areas of irregularity/sclerosis [Figure 1a and b]. She underwent resection through a posterior midline approach confirmed on the postoperative CT [Figure 1c and d]. She was asymptomatic within 6 postoperative weeks and remains symptom-free 2 years later.

Patient 2

A 16-year-old female presented with 2 years of low back pain and 3 months of pain radiating into her left hip refractory to NSAIDs and physical therapy. Comorbidities included recurrent migraines, major depressive disorder, obstructive sleep apnea, and restless leg syndrome. Her neurological examination was normal. The lumbar CT and MRI studies showed Bertolotti's syndrome on the left at the L5-S1 level characterized by an enlarged left L5 transverse process fused with the ilium and sacrum, with mild degeneration/sclerosis of the left L5 pars interarticularis [Figure 2a and b]. After surgical resection, confirmed on postoperative CT, she was intact and remained so 1 year postoperatively [Figure 2c and d].

DISCUSSION AND REVIEW OF RELEVANT LITERATURE

Diagnosis and pathophysiology of Bertolotti's syndrome

Bertolotti's syndrome is found in 10% of patients presenting with back and leg pain under 30 years of age. The biomechanics of LSTV is attributed to an alteration or reduction of movement between the transitional vertebra and the sacrum that can ultimately lead to pain from stress in the facet joint and/or is exacerbated by disc degeneration.\textsuperscript{[9]}

| Table 1: Castellvi classification schema of Bertolotti's syndrome by laterality (unilateral vs. bilateral) and morphological characteristics (size and anatomy involved) of the abnormal vertebral articulation. |
|----------------------------------------------------------------------------|
| **Castellvi classification system for lumbosacral transitional vertebra** |
| **Type** | **Characteristics** | **Specification** |
| I | Enlarged, dysplastic transverse process (>19 mm craniocaudal width) | Ia: Unilateral | Ib: Bilateral |
| II | Pseudoarticulation of the transverse process and sacrum with incomplete lumbarization/sacralization; enlargement of the transverse process w/pseudoarthrosis with a diarthrodial joint between itself and the sacrum | IIa: Unilateral | IIb: Bilateral |
| III | Transverse process completely fused with the sacrum with complete lumbarization/sacralization | IIIa: Unilateral | IIIb: Bilateral |
| IV | Type IIa pseudoarticulation on one side of vertebrae and Type IIIa fusion on the contralateral side |

**Figure 1:** Preoperative coronal computed tomography (CT) (a) and sagittal CT (b), both demonstrating a sacralized L5 with pseudoarthrosis from L5 to S1 and the ilium (white arrows). Postoperative coronal CT (c) and axial CT (d), indicating removal of the abnormal articulation (white arrows).

**Figure 2:** Preoperative coronal computed tomography (CT) (a) and sagittal CT (b), both demonstrating an extended left L5 transverse process fused with the ilium and sacrum (white arrows). Postoperative coronal CT (c) and axial CT (d), indicating removal of the abnormal articulation (white arrows).
Radiographic analysis

Radiographs, including flexion, extension views, and oblique views, confirm the diagnosis of Bertolotti’s syndrome. Both CT and MRI also demonstrate Bertolotti’s syndrome and also readily identify associated stenosis, osteophytes, and sclerosis adjacent to the articulation between the lumbar segment and ilium and/or sacrum.

Management of Bertolotti’s syndrome

Surgical resection of LSTV should be considered in patients presenting with intractable low back pain despite conservative treatment (e.g., physical therapy, nonsteroidal anti-inflammatory drugs, and localized anesthetic blocks in adults Figure 3).[7]

Outcomes of surgery with Bertolotti’s syndrome

There are few reports for patients under 18 years of age treated for Bertolotti’s syndrome [Table 2].[8] In three pediatric studies, two 17 years old experienced relief of their back and leg pain at 6 months and 1 year after surgery; in another, a 13 years old reported no improvement at 6-month follow-up; in a third, an 18 years old (in the same study) reported total alleviation of back pain 2 years after surgery.[3,4,8] Likely, Bertolotti’s syndrome is underdiagnosed in the pediatric population. Although surgical outcomes are generally positive, no randomized studies have documented their efficacy versus conservative nonsurgical treatment.

CONCLUSION

Bertolotti’s syndrome is seen in 10% of patients presenting with back and leg pain under 30 years of age.[16] While few surgical cases are reported in pediatric patients, removal of the abnormal transverse apophysis and disconnection from the lumbar spine/sacral ala should be considered as for those who fail conservative treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given her consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.
Table 2: Bertolotti’s syndrome surgical intervention and outcome studies.

| Study                  | Year | #Cases | Mean age (year) | Level/Type | Procedure                                                                 | Follow-up | Outcomes                                                                 |
|------------------------|------|--------|-----------------|------------|---------------------------------------------------------------------------|-----------|--------------------------------------------------------------------------|
| **Adolescents and adults** |      |        |                 |            |                                                                           |           |                                                                          |
| Jönsson et al. [8]     | 1989 | 11     | 39 (Range: 13–76) | n/a        | Resection of anomalous LSTV articulation                                 | 6 months–3 years | Seven patients pain resolved                                             |
|                        |      |        |                 |            |                                                                           |           | Two patients no change in pain                                           |
| Brault et al. [4]      | 2001 | 1      | 17              | L6-S1 Type Ila | Resection of the right L6 anomalous transverse process                    | 1 year   | Remission of LBP and leg pain w/no limitations in activity               |
| Babu et al. [3]        | 2017 | 2      | 17, 38          | L5-S1 Type Iib | Resection of LSTV utilizing O-arm                                         | 6 months, 9 months | Resolution of pain                                                        |
|                        |      |        |                 |            |                                                                           |           | Resolution of pain                                                        |
| **Adults**             |      |        |                 |            |                                                                           |           |                                                                          |
| Santavirta et al. [17] | 1993 | 16     | 34 (Range: 27–58) | L5-S1 Type Ia | 8: posterior-lateral L5-S1 fusion, 8: resection of LSTV articulation      | 2–17 years | 10 patients w/improved LBP                                               |
|                        |      |        |                 |            |                                                                           |           | Seven patients resolved LBP                                               |
|                        |      |        |                 |            |                                                                           |           | 11 of 13 patients w/continued sciatica                                   |
|                        |      |        |                 |            |                                                                           |           | Six patients underwent reoperation                                        |
| Abe et al. [1]         | 1997 | 1      | 37              | L5-S1 Type Iib | Extraforaminal decompression of the left L5 transverse process and bony  | 2 weeks, 1 year | Relief of LBP and leg pain; increased hypesthesia, numbness, and mild     |
|                        |      |        |                 |            | spur through extraperitoneal anterior approach                            |           | weakness                                                                  |
|                        |      |        |                 |            |                                                                           |           | Resolution of pain, hypesthesia, and numbness; no work limitations       |
| Ichihara et al. [6]    | 2004 | 1      | 34              | L5-S1 Type Iib | Resection of LSTV and nerve decompression through posterior approach      | 1 year    | Improved hip and leg pain                                               |
|                        |      |        |                 |            |                                                                           |           |                                                                          |
| Ugokwe et al. [20]     | 2008 | 1      | 48              | L5-S1 Type Iia | Minimally invasive resection of the left L5 transverse process and       | 6 weeks, 6 months | 10% improvement in pain                                                  |
|                        |      |        |                 |            | pseudoarticulation                                                       |           | 90% LBP and leg pain relief                                              |
| Weber et al. [21]      | 2010 | 1      | 53              | L5-S1 Type Iia | Lateral foraminal and extraforaminal nerve decompression through posterior | 1 year    | No LBP or radicular pain                                                 |
|                        |      |        |                 |            | approach                                                                  |           |                                                                          |
| Shibayama et al. [18]  | 2011 | 1      | 46              | L6-S1 Type Iia | Extraforaminal decompression of the right L6 body, transverse process,   | 30 months | Improved LBP, sciatica, and returned to work                              |
|                        |      |        |                 |            | and upper sacral ala                                                      |           |                                                                          |
| Miyoshi et al. [15]    | 2011 | 1      | 29              | L5-S1 Type Iib | Resection of the right LSTV osteophytes/pseudoarticulation               | 1 month, 1 year | Improved R-sided leg and buttock pain                                   |
|                        |      |        |                 |            |                                                                           |           | R-sided pain resolved                                                     |
| Kikuchi et al. [11]    | 2013 | 2      | 70, 53          | L5-S1 Type Iia | Decompression and LSTV resection through anterior approach               | 1 year, 1 year | LBP and leg pain relieved and functionally improved                       |
| Malham et al. [13]     | 2013 | 2      | 27, 49          | L5-S1 Type Iia | Resection of LSTV through anterior, retroperitoneal approach             | Patient 1 (27 years) | Improved LBP and return to work                                          |
|                        |      |        |                 |            |                                                                           | Patient 2 (49 years) | No exercise limitations                                                   |
|                        |      |        |                 |            |                                                                           |           | Improved LBP and return to part-time work No limitations with low-impact  |
|                        |      |        |                 |            |                                                                           |           | exercise                                                                  |

(Contd.)
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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Abe E, Sato K, Shimada Y, Okada K, Yan K, Mizutani Y, et al. Anterior decompression of foraminal stenosis below a lumbosacral transitional vertebra. A case report. Spine (Phila Pa 1976) 1997;22:823-6.

2. Adams R, Herrera-Nicol S, Jenkins AL 3rd. Surgical treatment of a rare presentation of Bertolotti’s syndrome from castellvi type IV lumbosacral transitional vertebra: Case report and review of the literature. J Neurol Surg Rep 2018;79:e70-e74.

3. Babu H, Lagman C, Kim TT, Grode M, Johnson JP, Drazin D, et al. Intraoperative navigation-guided resection of anomalous transverse processes in patients with Bertolotti’s syndrome. Surg Neurol Int 2017;8:236.

4. Brault JS, Smith J, Currier BL. Partial lumbosacral transitional vertebra resection for contralateral facetogenic pain. Spine (Phila Pa 1976) 2001;26:226-9.

5. Holm EK, Bünger C, Foldager CB. Symptomatic lumbosacral transitional vertebra: A review of the current literature and clinical outcomes following steroid injection or surgical intervention. SICOT J 2017;3:71.

6. Ichihara K, Taguchi T, Hashida T, Ochi Y, Murakami T, Kawai S, et al. The treatment of far-out foraminal stenosis below a transitional vertebra: A report of two cases. J Spinal Disord Tech 2004;17:154-7.

7. Jain A, Agarwal A, Jain S, Shamshery C. Bertolotti syndrome: A diagnostic and management dilemma for pain physicians. Korean J Pain 2013;26:368-73.

8. Jönsson B, Strömqvist B, Egund N. Anomalous lumbosacral articulations and low-back pain. Evaluation and treatment. Spine (Phila Pa 1976) 1989;14:831-4.

9. Ju CI, Kim SW, Kim JG, Lee SM, Shin H, Lee HY, et al. Decompressive L5 transverse processectomy for Bertolotti's syndrome: A preliminary study. Pain Physician 2017;20:E923-E932.

10. Kapetanakis S, Chaniotakis C, Paraskevopoulos C, Pavlidis P. An unusual case report of Bertolotti’s syndrome: Extraforaminal stenosis and L5 unilateral root compression (Castellvi type III an LSTV). J Orthop Case Rep 2017;7:9-12.

11. Kikuchi K, Abe E, Miyakoshi N, Kobayashi T, Abe T, Hongo M, et al. Anterior decompression for far-out syndrome below a transitional vertebra: A report of two cases. Spine J 2013;13:e21-5.

12. Li Y, Lubelski D, Abdullah KG, Mroz TE, Steinmetz MP. Minimally invasive tubular resection of the anomalous transverse process in patients with Bertolotti’s syndrome: Presented at the 2013 joint spine section meeting. Clinical article. J Neurosurg Spine 2014;20:283-90.

13. Malham GM, Limb RJ, Claydon MH, Brazenor GA. Anterior pseudoarthrectomy for symptomatic Bertolotti’s syndrome.
14. Manmohan S, Dzulkarnain A, Azlin ZA, Fazir M. Bertolotti’s syndrome: A commonly missed cause of back pain in young patients. Malays Fam Physician 2015;10:55-8.

15. Miyoshi Y, Yasuhara T, Date I. Posterior decompression of far-out foraminal stenosis caused by a lumbosacral transitional vertebra case report. Neurol Med Chir (Tokyo) 2011;51:153-6.

16. Quinlan JF, Duke D, Eustace S. Bertolotti’s syndrome. A cause of back pain in young people. J Bone Joint Surg Br 2006;88:1183-6.

17. Santavirta S, Tallroth K, Ylinen P, Suoranta H. Surgical treatment of Bertolotti’s syndrome. Follow-up of 16 patients. Arch Orthop Trauma Surg 1993;112:82-7.

18. Shibayama M, Ito F, Miura Y, Nakamura S, Ikeda S, Fujiwara K, et al. Unsuspected reason for sciatica in Bertolotti’s syndrome. J Bone Joint Surg Br 2011;93:705-7.

19. Takata Y, Sakai T, Higashino K, Goda Y, Mineta K, Sugiura K, et al. Minimally invasive microendoscopic resection of the transverse process for treatment of low back pain with Bertolotti’s syndrome. Case Rep Orthop 2014;2014:613971.

20. Ugokwe KT, Chen TL, Klineberg E, Steinmetz MP. Minimally invasive surgical treatment of Bertolotti’s syndrome: Case report. Neurosurgery 2008;62:ONSE454-5.

21. Weber J, Ernestus RI. Transitional lumbosacral segment with unilateral transverse process anomaly (Castellvi type 2A) resulting in extraforaminal impingement of the spinal nerve: A pathoanatomical study of four specimens and report of two clinical cases. Neurosurg Rev 2010;34:143-50.