Occipitopexy as a Fusionless Solution for Dropped Head Syndrome

A Case Report

Mazda Farshad, MD, MPH, Marco D. Burkhard, MD, and José M. Spirig, MD

Investigation performed at Department of Orthopaedic Surgery, Balgrist University Hospital, University of Zurich, Zurich, Switzerland

Abstract

Case: A 68-year-old woman suffered from an irradiation-induced dropped head syndrome (DHS). Fusion surgery was vehemently rejected by the patient. A new surgical method, avoiding fusion, was invented and performed to treat her DHS. This novel surgical technique of "occipitopexy"—a ligamentous fixation of the occiput to the upper thoracic spine—is described in detail. One year postoperatively, the patient was very satisfied, able to maintain a horizontal gaze, and rotate her head 20° to each side.

Conclusion: This is the first report describing "occipitopexy" as an alternative to cervicothoracic fusion for patients with flexible DHS.

Dropped head syndrome (DHS) is a rare disorder characterized by a gradual weakening of the cervical musculature, causing an inability to extend the neck and resulting in a debilitating curvature of the cervical spine. In contrast to other kyphotic cervical spine disorders such as ankylosing spondylitis, the "chin-on-chest deformity" in DHS is often flexible, and an upright posture can be achieved with passive neck extension[1]. Various etiologies have been reported, including neuromuscular disorders (i.e., myasthenia gravis, amyotrophic lateral sclerosis, and Parkinson's disease) and postsurgical and irradiation therapies[2,3]. In a subgroup of DHS defined as isolated neck extensor myopathy[4], no other underlying conditions can be found. Besides potential neurologic impairments, affected individuals may be severely restricted in ambulation and activities of daily living, including eating and social interaction[5]. First-line management for DHS comprises

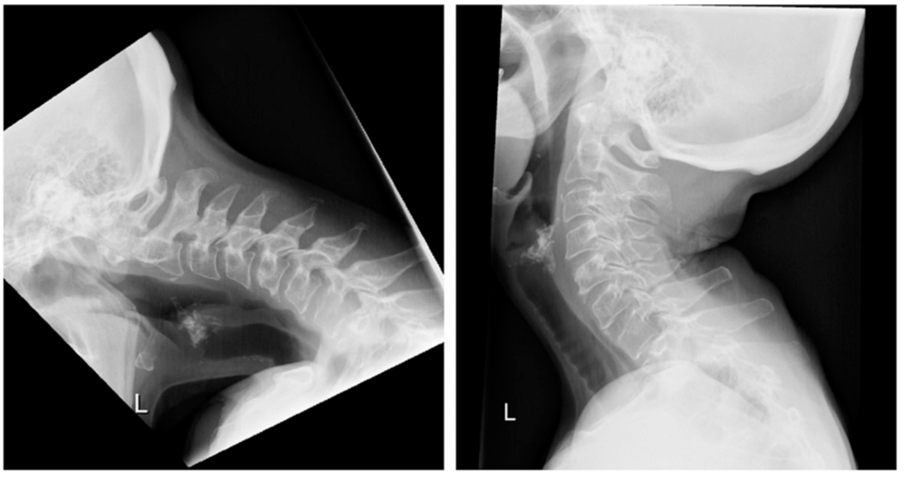

Fig. 1

Preoperative radiographs showing the dropped head in the neutral position and full passive reducibility of dropped head deformity with moderate degenerative changes.

Disclosure: The **Disclosure of Potential Conflicts of Interest** forms are provided with the online version of the article (http://links.lww.com/JBJSCC/B580).

Keywords: occipitopexy, dropped head, neck extensor, cervical fusion, Hodgkin lymphoma

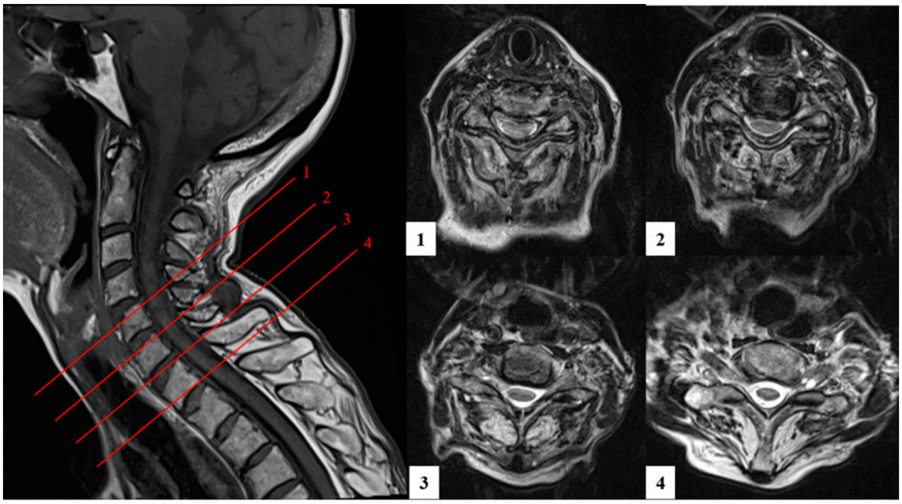

Fig. 2
Preoperative magnetic resonance imaging (T2-weighted) showing the irradiation-induced atrophy, scarring, and fatty infiltration of the paraspinal muscles (axial) and mild spinal stenosis but no signs of myelopathy. Slice numbers on the sagittal view correspond to the figure subnumbers.

therapy of the underlying condition with concomitant physical therapy and/or the use of a cervical collar[3]. If conservative management fails, only a few surgical options are available. Long-construct cervical fusion from posterior or combined anteroposterior fusions and cervical pedicle subtraction osteotomies have been described at the level of case reports and case series[5-7].

To date, no motion-preserving alternatives have been described to restore cervical sagittal balance. In this case report, we report the first fusionless reconstruction of cervical lordosis using a tendon-allograft fixation construct from the occiput to T1/2 for a patient with irradiation-induced DHS.

The patient was informed that data concerning the case would be submitted for publication, and she provided consent.

Case Report

A 68-year-old woman initially presented with a passively correctable “chin-on-chest deformity” and an inability to hold her head upright without external aid since several years. Her medical history was significant for Hodgkin lymphoma of the neck, which had been treated with radiation therapy 35 years earlier. No underlying neuromuscular disorder or other causes for the weakened cervical muscles, other than the history of irradiation, could be identified. The patient reported the development of DHS starting 24 years after the radiation therapy. The patient required her hands as a chin support to maintain a horizontal gaze; otherwise, she denied any pain and presented neither neurological deficits nor clinical signs of myelopathy. The radiographs documented age-related degenerative changes but no other abnormalities (Fig. 1). The magnetic resonance imaging of the cervical spine showed no neural compression (Fig. 2). However, irradiation had likely induced atrophy, scarring, and fatty infiltration of the paraspinal muscles, along with edematous changes in the cranial portion of the trapezius and scalenus muscles.

The patient did not tolerate head support with orthosis. Cervicothoracic fusion surgery was discussed but vehemently refused by the patient. With continuous worsening of cervical and thoracic hyperkyphosis (Fig. 3), the patient aimed for a definite solution for her DHS but still refused any fusion

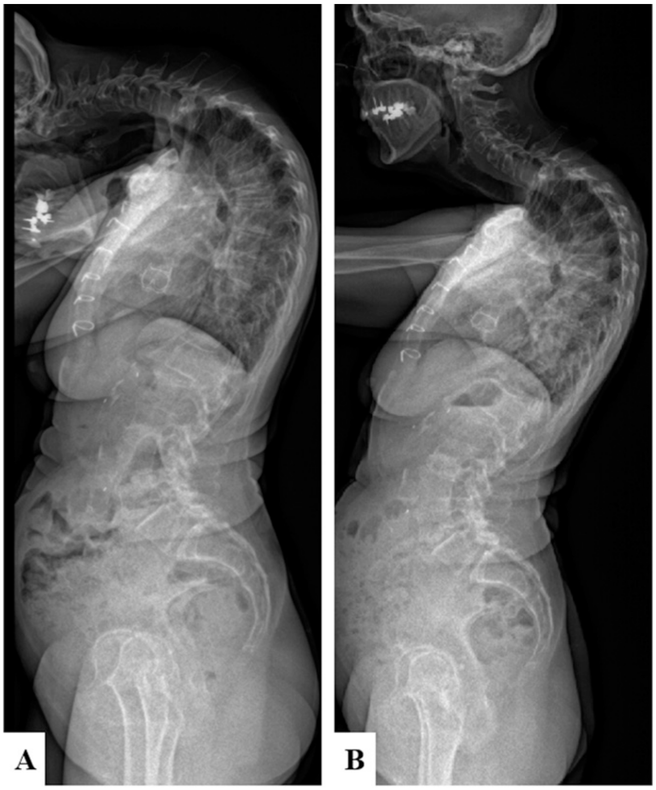

Fig. 3
Fig. 3-A Preoperative and **Fig. 3-B** 1-year postoperative standing long cassette radiographs.

options. As a consequence, the so-called "occipitopexy" was developed as a reconstruction of the cervical lordosis. The surgical procedure was exercised in a cadaver specimen to test feasibility before being offered to the patient. The patient was aware of the experimental character of this procedure and gave informed consent.

Surgical Technique

The patient was positioned prone and the head positioned with a horizontal gaze using a Mayfield scull clamp fixation (Fig. 4-A). A midline posterior skin incision was used to expose the occiput, the spinous processes, and laminae from C0 to T2. To avoid dorsal protrusion of a traction cable ("bowing") from

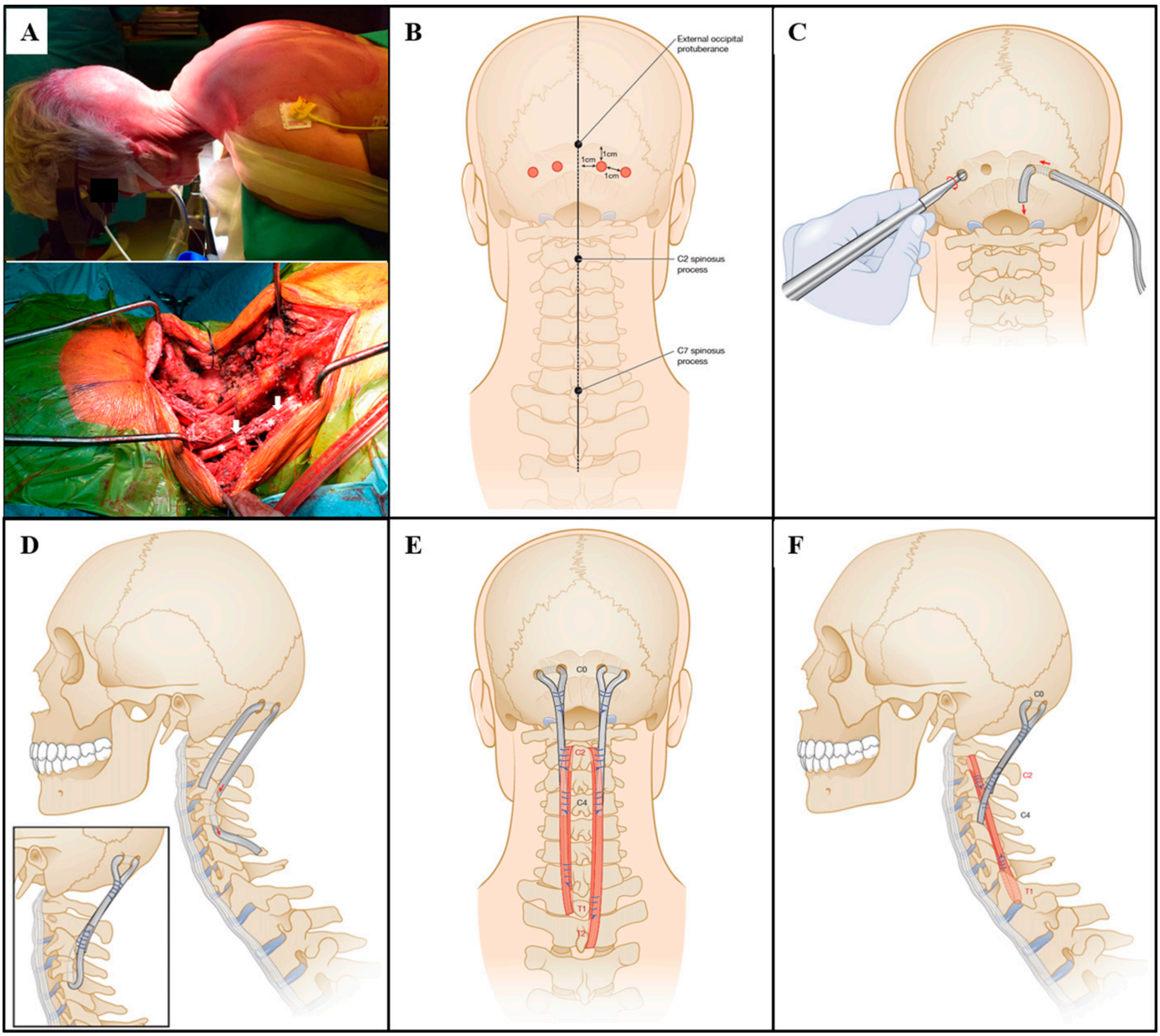

Fig. 4

Schematic illustrations and intraoperative photographs of the occipitopexy. **Fig. 4-A** Intraoperative photographs of the patient positioning in the Mayfield clamp and the situs with the allograft tendon (asterisks) and sublaminar polyester (Nile) band construct (arrows). **Fig. 4-B** Landmarks: The occiput trepanations were positioned 1 cm inferior of the superior nuchal line and 1 cm lateral to the midline. The lateral holes were accordingly set 1 cm lateral to the medial holes. **Fig. 4-C** After trepanation, the intracranial dura was separated from the occiput with a dissector, and allograft tendons and Nile bands were pulled through the trepanation holes from lateral to medial. **Fig. 4-D** Caudally, the tendons were passed around the lamina of C4. **Fig. 4-E** Further allograft tendons and Nile bands were then threaded from the lamina of C2 to T1 on the left and to T2 on the right. **Fig. 4-F** By using the overlapping tendon construct, dorsal protrusion of a traction table could be avoided.

Fig. 5
Global balance. **Fig. 5-A** Preoperative. **Fig. 5-B** One year postoperative.

C0 to T1 above the skin level, a fibrous fixation from the occiput to C4 and from C2 to T1 was aimed.

Bilateral trepanation of the occiput with two 4-mm burr holes on each side was performed. The burr holes were placed 1 cm below the superior nuchal line. The medial trepanations were placed at 1-cm distance from the midline and the lateral trepanations 1 cm lateral from the medial holes (Fig. 4-B). The intracranial dura was separated from the occiput with a dissector. On each side, a gracilis tendon allograft together with a reinforcing sublaminar band consisting of polyethylene terephthalate, namely a Nile band (K2M, Stryker), was threaded from the lateral to the medial hole (Fig. 4-C).

The lamina of cervical vertebra C4 was prepared for tendon and band passage by removing the attachments of the ligamentum flavum on both the cephalad and caudal ends of the lamina, creating interlaminar windows at C3/4 and C4/5 on both sides. The distal ends of the tendons and the Nile bands were then carefully passed underneath the C4 lamina on both sides from cephalad to caudal. Using a rongeur, the tips of both the tendons and the Nile bands were then pulled upward through the interlaminar windows at C4/5. The tendons and Nile bands were further advanced to form loops on each end, one passing through the occiput and another passing around the lamina of C4. The bilateral traction cables from C0 to C4 so formed were then tightened and fixated with No. 2 Ethibond sutures (Ethicon Johnson & Johnson) in a 2-cm-long Krackow suture (Fig. 4-D)[8]. Similarly, the bilateral traction cables between the lamina of C2 and T1 on the left and C2 and T2 on the right were constructed (Fig. 4-E). Different thoracic fixation points (T1 and T2) on each side were chosen to reduce the risk of lamina tear-out fracture. Traction on the tendons was applied before suturing the tendon ends. The crossing points between the upper (C0-C4) and the lower (C2-T1/T2) ligamentous constructs were further sutured at the level of C4 to

Fig. 6
Postoperative range of motion. **Fig. 6-A** Flexion/extension of 0-0-10. Downward gaze possible by forward bending. **Fig. 6-B** Side rotation right/left 20-0-20.

have a connected traction cable from C0 to the thoracic area (Fig. 4-F). The nuchal fascia was then exposed to tighten it lengthwise by oversewing redundant folds over the now lordotic cervical spine. The fascia, subcutis, and skin were closed in the usual manner.

Postoperative Course

The cervical spine was immobilized in a hard collar for 4 months after surgery, followed by a mild mobilization of the neck after 4 months. Unrestricted range of motion was allowed after 6 months, followed by further mobilization and strengthening exercises. One year after surgery, the patient is very satisfied, has an upright posture with a horizontal gaze, and does not need to use her hands to hold her head up (Fig. 5). She is pain-free, except for some occasional and tolerable muscular tensions of the neck, and does not need any analgesics. She is able to rotate her head 20° in each direction and extend her neck 10° (Fig. 6). The irradiation-induced muscular atrophy might have limited further improvement of the range of motion 6 months postoperatively.

Discussion

To the best of our knowledge, this is the first description of a fusionless surgical correction of a DHS worldwide. The occipitopexy—an innovative technique including a fixation from C0 to T1/T2 using tendon allograft and synthetic bands—led to cervical lordosis restoration and pain-free range of motion of 10° in extension and 20° in rotation on each side.

Development of DHS after irradiation of Hodgkin lymphoma has been reported by a few authors[7,9,10]. Furby et al.[9] reported a case series of 6 patients developing DHS 5 to 30 years after radiation treatment. A systematic review by Drain et al.[6] yielded a positive clinical outcome in 15 of 16 reported cases undergoing fusion surgery for DHS. Most of the reported patients underwent a fusion from C2 to the upper thoracic vertebrae. Only 4 cases in the literature underwent an occipitocervicothoracic fusion[11-14]. Theoretically, a spinal fusion from C2 to T1 would preserve motion between the occiput, atlas, and axis, which in healthy subjects is 14° of flexion/extension, 2° of lateral flexion, and 75° of axial rotation[15]. However, data about occipitoatlantoaxial motion after subaxial fusion are limited. Petheram et al. have reported poor outcome after subaxial fusion of a patient with DHS with grossly limited neck movements with 5° of extension[16]. The postoperative loss of mobility of the cervical spine after fusion surgery may place patients at risk of falls because of the inability to see the ground in front of the feet[16]. In this context, a ligamentous fixation reaching to C2 instead of the occiput was considered in our case to theoretically allow more motion in the craniocervical junction. However, this would have put the whole ligamentous construct into more stress and increase the risk of failure because of the more anterior fixation points of C2 compared with C0.

Another concern for fusion surgery is screw hold in osteoporotic bone. In the case described here, previous radiation therapy added a risk factor for osteoporosis[16-18]. The surgical technique used does not require bone fusion and avoids complications at the screw–bone interface. However, whether the ligamentous construction technique reported here will maintain the upright posture in the long term remains unknown. Only limited parallels to other types of ligament reconstructions can be drawn because the pathogenesis and biomechanical considerations of irradiation-induced DHS are unique. If the reconstruction described here fails, graft removal and long-construct cervicothoracic fusion would be proposed as a rescue option.

In sum, with only 1 case described, no superiority can be claimed for the fusionless occipitopexy vs. the state-of-the-art fusion-based solution for surgical treatment of DHS. The occipitopexy could be regarded as a considerable alternative to a cervicothoracic fusion in patients with flexible DHS. Success at 1 year may indicate the value of this surgical technique, but the long-term results remain to be observed in the future.

Conclusion

In this case report, the occipitopexy—a ligamentous fixation of the occiput to the upper thoracic spine—was introduced as a fusionless solution to DHS. The satisfactory clinical and radiographic results after a year indicate that this procedure could be a worthwhile alternative to cervical thoracic fusion in patients with flexible DHS. ■

Mazda Farshad, MD, MPH[1]
Marco D. Burkhard, MD[1]
José M. Spirig, MD[1]

[1]Department of Orthopaedic Surgery, Balgrist University Hospital, University of Zurich, Zurich, Switzerland

E-mail address for M. Farshad: mazda.farshad@balgrist.ch

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