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A Novel Treatment of Pediatric Atlanto-Occipital Dislocation with Nonfusion Using Muscle-Preserving Temporary Internal Fixation of C0-C2: Case Series and Technical Note

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Abstract

Study Design Case series with surgical technical note.

Objectives This article reports experiences and results of muscle-preserving temporary C0-C2 fixation for the treatment of atlanto-occipital dislocation (AOD).

Methods AOD is a rare injury caused by high-energy trauma, occurring in less than 1% of pediatric trauma patients. Recommended treatment is C0-C2 fusion which, however, will result in significant loss of mobility in the craniocervical junction (CCJ), especially C1-C2 rotation. An alternative approach, with the ability of preserving mobility in the C1-C2 segment, is a temporary fixation that allows the ligaments to heal, after which the implants can be removed to regain function in the CCJ joints. By using a muscle-preserving approach and navigation for the C2 screws, a relatively atraumatic fixation of the CCJ can be achieved with motion recovery after implant removal.

Results We present two cases of AOD treated with temporary fixation. A 12-year-old boy involved in a frontal car collision, as a strapped back seat passenger, was treated with temporary C0-C2 fixation for 10 months. Follow-up at 11 months after implant removal included clinical evaluation, computed tomography, magnetic resonance imaging (MRI), and flexion-extension X-rays. He was free of symptoms at follow-up. The CCJ was radiographically stable and he had 45 degrees of C1-C2 rotation.

A 7-year-old girl was hit by a car as she got off a bus. She was treated with temporary fixation for 4 months after which the implant was removed. Follow-up at 8 years included clinical evaluation and MRI in rotation. She was free of symptoms. The ligaments of the CCJ appeared normal and her C1-C2 rotation was 30 degrees.

Conclusion C0-C2 fixation without fusion allows the CCJ ligaments to heal in pediatric AOD. By removing the implants after ligament healing, rotation in the C1-C2 segment

Keywords
- navigation
- atlanto-occipital dislocation
- occipitocervical dissociation
- craniocervical dissociation
- muscle preservation
- motion preservation

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can be regained without subsequent instability. Both our patients tolerated the treatment well and were free of symptoms at follow-up.
By using minimally invasive muscle-preserving technique and navigation, temporary fixation of the CCJ can be achieved with minimal damage to the soft tissues allowing recovery of almost normal function after implant removal.

Introduction

Traumatic atlanto-occipital dislocation (AOD), also called occipitocervical dissociation or craniocervical dissociation, is a very rare injury, associated with high-energy trauma, where the ligaments of the craniocervical junction (CCJ) are torn and the head is separated from the cervical spine. The condition was first described by Blackwood in 1908 as a case report.[1] The mortality at the scene of the accident is probably high, but there are some reports of survivors mainly in the pediatric population with no more than level III evidence for diagnosis and treatment. A systematic review from 2019 describes 144 cases of pediatric AOD in retrospective studies and case reports available through PubMed, of whom 96 survived the injury and 86 underwent an occipitocervical fusion (OCF).[2] In a long-term evaluation at a level 1 trauma center, the incidence of AOD was 0.6% of 5,337 spine trauma cases.[3]

Diagnosing AOD may be challenging. In high-energy trauma victims, the primary radiographic investigation is usually computed tomography (CT). When assessing the CCJ, common parameters are the condyle-C1 interval (CCI), basion-dens interval (BDI), basion-axis interval, and the powers ratio (►**Fig. 1**). Of these parameters, the most accurate parameter is CCI, especially in the pediatric population. Threshold values for CCI, indicating AOD, are greater than 1.5 mm for adults and greater than 4 mm for children.[4–6] Threshold values for BDI are greater than 10 mm in adults and greater than 12 mm in children. For powers ratio, less than 0.9 is considered normal whereas greater than 1 indicates suspicion for AOD.[7–9] In a late review, CCI was used in 23% of cases as indication for fusion, BDI in 14%, and powers ratio in 14%.[2] Magnetic resonance imaging (MRI) is an important modality for evaluating the alar and apical ligaments and the tectorial membrane. Other tell-tale signs of AOD are retroclival hematoma, and hematoma in the C0-C1 or C1-C2 joints. Final diagnosis is usually established by a combination of CT and MRI.[5,10–12] A traction test under anesthesia using fluoroscopy may also be used; if the C0-C1 is distended more than 2 mm OCF is advised.[10]

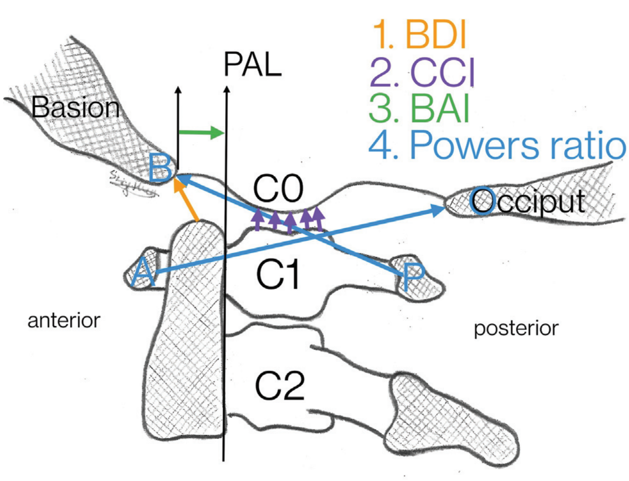

Fig. 1 Yellow (1): basion-dens interval (BDI). Purple (2): condyle-C1 interval (CCI). Green (3): basion-axis interval (BAI) (PAL = posterior axial line). Blue (4): powers ratio (BP/OA).

As the instability caused by AOD is potentially life-threatening, the recommended treatment is OCF.[2,10,13–17] Large variation of OCF types have been used over time, from sublaminar wires to all types of C2 screw techniques. Today, the most common treatment involves occipital plate and C2 pedicle screws.[2,5,17–23] Temporary fixation, pending surgery, using hard collar or halo-vest works well,[5,11,14,19] whereas skull traction is contraindicated as the risk increased of neurologic injury is up to 10%.[5,13,24] A major drawback of OCF is stiffness. C1-C2 fusion will eliminate approximately 50% of total cervical rotation, whereas locking C0-C1 will not only affect cervical extension and flexion, but may also add to dysphagia, and probably also increase the risk of premature disk degeneration at levels below. Thus, a temporary stabilization, allowing safe fixation while the soft tissues heal, followed by implant removal and regained mobility, would be beneficial. Such a technique would also have to preserve muscle function in the CCJ to regain meaningful function. We have applied temporary internal C0-C2 fixation via muscle-preserving approach and navigation in two cases. Permission to perform this follow-up was approved by the Swedish Ethical Review Authority (registration number: 2020–03097).

Case 1

A 12-year-old boy was a belted backseat passenger in a high-speed frontal car collision, a multitrauma with injury to abdomen with intestine involvement requiring surgery, a Chance fracture of L3, and AOD. The patient was neurologically intact with severe neck pain apart from his other injuries. The initial CT showed no fractures in the cervical spine, but hematoma posterior to the clivus, BDI 10 mm, CCI 5.5 mm, and subluxation C0-C1 suggesting a ligamentous injury of the CCJ (►**Fig. 2**). MRI revealed rupture of the tectorial membrane, and rupture and elongation of the alar ligaments (►**Fig. 2**). The patient was operated with temporary C0-C2 fixation via muscle-preserving approach and navigation (►**Fig. 3**). The

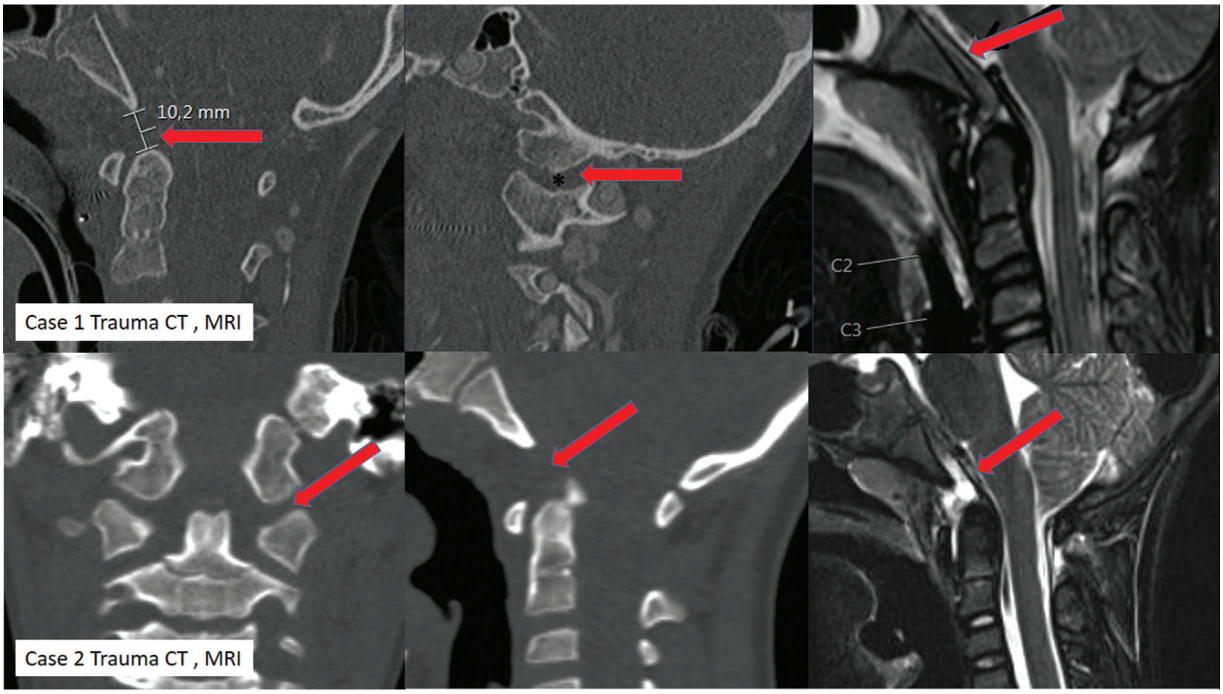

Fig. 2 Trauma computed tomography (CT) and magnetic resonance imaging (MRI) showing increase in condyle-C1 interval (CCI), basion-dens interval (BDI), and retroclival hematoma in cases 1 and 2.

implants were removed at 10 months, and after that MRI confirmed normal appearance of the soft tissues of the CCJ. Follow-up was at 3 and 11 months after implant removal. The patient was free of symptoms, had no neck pain, and is fully active. To assess stability and motion, CT in rotation, flexion-extension radiographs, and MRI were obtained. CT showed normal CCI and BDI, and 44 degrees of C1-C2 rotation (22 degrees dexter, 22 degrees sinister) at follow-up at 3 months. At 11-month follow-up, CT in rotation showed 39 degrees of C1-C2 rotation (21 degrees dexter, 18 degrees sinister). MRI showed good visualization of alar ligaments, assessed as radiologically intact, and the tectorial membrane was at normal position. Flexion-extension radiographs showed no excess C0-C1 movement.

Case 2

A 7-year-old girl was hit by a car as she got off a bus. Apart from AOD she sustained a head concussion, an undisplaced acetabular fracture, and a liver contusion. She was initially a little lethargic, but awake and neurologically intact. CT revealed increase in BDI and CCI, an asymmetrical widening

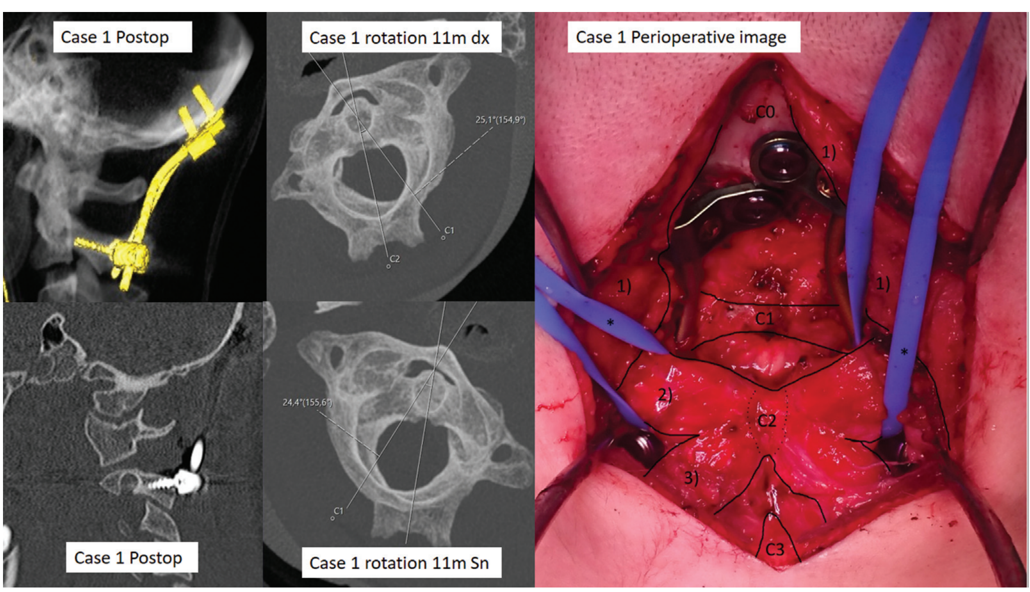

Fig. 3 Postop computed tomography (CT) showing screws and plate. Rotation at the 11-month follow-up. Perioperative image with (1) semispinalis capitis muscle, (2) rectus capitis posterior major and obliquus capitis inferior muscle, and (3) semispinalis cervicis muscle.

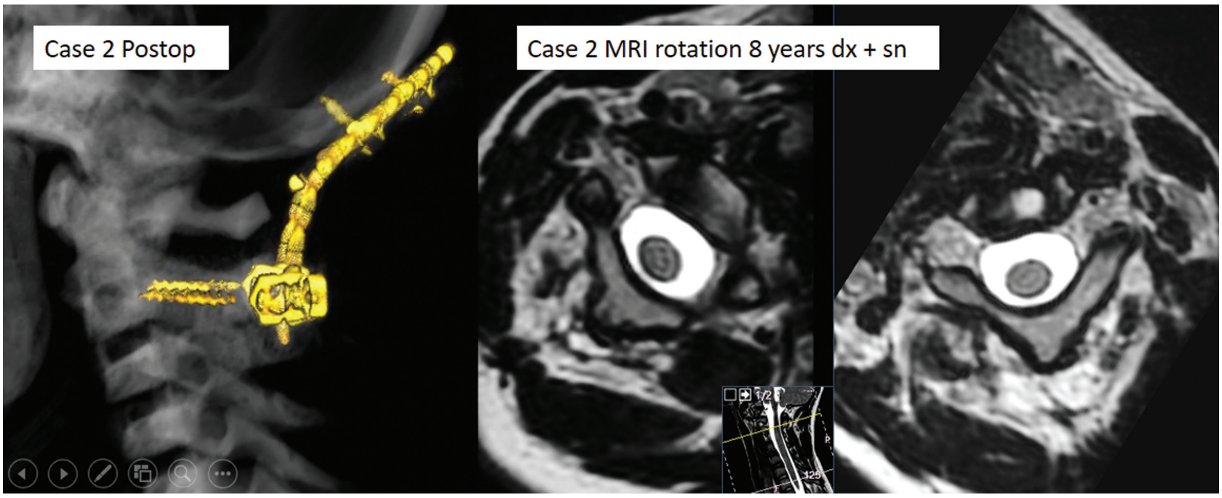

Fig. 4 Case 2: Postop computed tomography (CT) and magnetic resonance imaging (MRI) at the 8-year follow-up in rotation dexter (dx) and sinister (sn).

of the neurocentral synostosis of C2, displacement of the tip of the odontoid, and a retroclival hematoma. MRI showed a rupture of the tectorial membrane. A temporary C0-C2 fixation was performed with implant removal after 4 months. A long-term follow-up, including MRI in rotation, was made after 8 years. At this time the patient had no cervical symptoms and had a clinically normal range of cervical motion. An MRI of C1-C2 showed that the alar ligaments and tectorial membrane were normal and rotation MRI of the CCJ showed 30 degrees of motion in C1-C2 segment (9 degrees dexter, 21 degrees sinister), shown in ►**Fig. 4**.

Surgical technique: Navigated C0-C2 muscle-preserving fixation.

The main goal is to achieve a temporary rigid fixation from the occiput to the C2-vertebra without major injury of the C2 muscles.

The following six steps are made during the procedure: 1st step: To access the deep muscles of the neck with midline approach. 2nd step: To make space for C0 plates and screws with partial muscle removal. 3rd step: Make space for attachment of navigation reference on C2 spinal process without injury of muscles. 4th step: C2 pedicle screws placement using navigated probe or drill. 5th step: Tunneling of the rods under the muscles. 6th and final step: Attachment of C0 plates.

1st step: A midline posterior skin incision is made from the occiput to the C3 spinal process to provide access from the occipital protuberancia all the way to the C3 vertebra. Then, splitting of the nuche ligament is performed from occiput to C3 spinal process. Once the nuche ligament is split, one can easily dissect the connective tissue between nuche and the C2 spinal process without injury of the superficial muscles of the neck. The muscles are then moved laterally as shown in ►**Supplementary Fig. S1** (online only).

2nd step: Access to the occipital protuberancia is made by exposing the midline portion of occiput and approximately 1.5 to 2 cm laterally. This means removal of the insertion of the rectus capitis posterior minor muscle and partial removal of semispinalis capitis muscle. The goal is to make enough space for occipital plates and rods as shown in ►**Supplementary Fig. S2**, (online only); then, one continues dissection deep toward the C1 arch and the posterior tubercle of atlas to remove the origin of rectus capitis posterior minor muscle approximately 1 cm laterally from midline. Once C1 arch is exposed, one can easily access the space between C1 arch and the C2 spinal process and exposure of the epidural membrane C1-C2 as shown in ►**Supplementary Fig. S2**, (online only).

3rd step: Attachment of navigation reference to C2 is made by access of the interspinal space of C2-C3 and free cranial margin of C2. A midline dissection of C2-C3 will make space for the navigation reference which is attached to C2 spinal process as shown in ►**Supplementary Fig. S3** (online only).

4th step: O-arm scan is now performed with reference on C2 spinal process. Correct entry point of C2 pedicles are now located by using navigation and exposing the bony entry point on C2 with sharp neural dissector. Navigated pedicle probe is helpful due to the small pedicles of pediatric C2 pedicles. Screws are then placed without too much injury of muscle tissue and attachments. The rectus capitis major, obliquus inferior, and the cervicis spinalis muscles are attached to the C2 spinal process, in some cases they can be split without too much injury of the muscle.

5th step: Rod placement is made by first making a tunnel under the rectus capitis posterior major muscle and the obliquus capitis inferior muscle. This is done by a sharp neural dissector from the screws in following the lamina of C2 in direction of C1 midline. This needs to be done with care, shown in ►**Supplementary Fig. S4** (online only) by the blue rubber bands. Once rods are measured they can be inserted in craniocaudal direction under the muscles and attached to C2 screws.

6th step: Final step is adding the occipital plate to the rods and fixations of plates in midline of the occiput as shown in ►**Supplementary Fig. S4** (online only).

Discussion

Approaches to the posterior cervical spine using the intermuscular planes and muscle preservation is well documented and described in the subaxial cervical spine and CCJ.[25,26] This muscle-preserving CCJ approach starts with removal of the origin and insertion of the capitis rectus minor muscle to access the C1 tubercle and occiput. Also, a very small portion of the insertion of the semispinalis capitis muscle is removed to gain access to the occiput when attaching the occipital plate. It seems that lack of the capitis rectus minor muscle and a small portion of semispinalis capitis is compensated for by the rest of the CCJ muscles. Using navigation and the intermuscular planes to gain access to C2 makes it possible to find the C2 entry points without removing muscles from C2. Using this concept to keep as much as possible of the soft tissues in children with AOD, seems favorable to preserve functionality of the segment. There is always some degree of fibrotic tissue, however, that reduce some of the movement but not as much as an overall fusion. The segment maintains 30 to 45 degrees of motion after implant removal, even after 8 years (case 2). We think that this preserved motion could be protective against degeneration of the C2-C3 segment. We have not seen any increased instability in the segment after implant removal in the short term or in the long term. This suggests that the ligaments of the CCJ are sufficiently healed, and this is confirmed by flexion and extension radiograph, CT, and MRI. The healing potential seems so good that we could see intact appearance of the alar ligament and the tectorial membrane in both cases on MRI (►**Fig. 5**). We believe that the doctrine that ligamentous injury in pediatric populations must be treated with fusion must be questioned.

Conclusion

We have shown that temporary internal fixation of C0-C2 allows the CCJ ligaments to heal. Four to 10 months of stabilization are sufficient to heal the ligamentous injury to such degree that there are no signs of instability at all follow-ups. Clinically, the patients were doing well with very little neck pain. We strongly believe this is an alternative treatment to AOD in pediatric population.

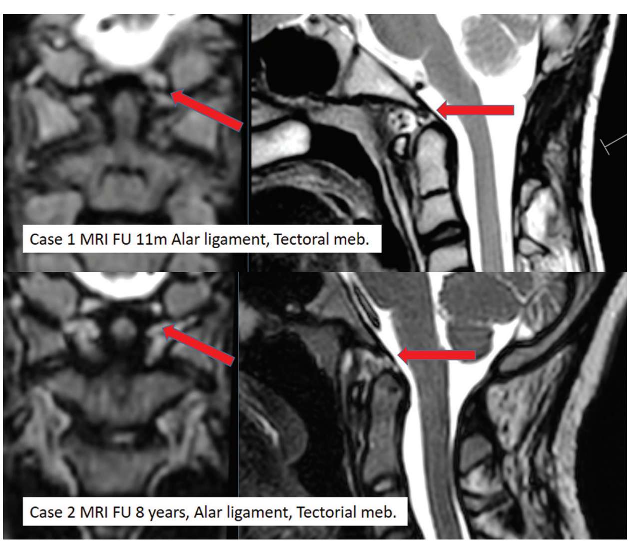

Fig. 5 Alar ligament and tectorial membrane during follow-up, are intact.

Authors' Contributions

Conception and design: M.H., C.O.
Surgical technique: M.H., C.O.,
Drafting the article: M.H., L.S.
Illustrations: L.S.
Photos: M.H.
Critically revising the article: C.O., A.J.
Study supervision: C.O.

Ethical Permission

Permission to perform this follow-up was approved by the Swedish Ethical Review Authority (registration number: 2020–03097).
All included patients consented to procedure.

Funding

Institutional funding: The funder played no part in study design; collection, management, analysis, and interpretation of data; writing of the report; and the decision to submit the report for publication.

Conflict of Interest

None declared.

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