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Case Report

C-arm Free O-arm Navigated Posterior Atlantoaxial Fixation in Down Syndrome: A Technical Note

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The surgical treatment of pediatric atlantoaxial subluxation (AAS) in Down syndrome (DS) remains technically challenging due to radiation exposure and complications such as vertebral artery injury and nonunion. The established treatment is fixation with a C1 lateral mass screw and C2 pedicle screw (modified Goel technique). However, this technique requires fluoroscopy for C1 screw insertion. To avoid exposing the operating team to radiation we present here a new C-arm free O-arm navigated surgical procedure for pediatric AAS in DS. A 5-year-old male DS patient had neck pain and unsteady gait. Radiograms showed AAS with an atlantodental interval of 10 mm, and irreducible subluxation on extension. CT scan showed Os odontoideum and AAS. MRI demonstrated spinal cord compression between the C1 posterior arch and odontoid process. We performed a C-arm free O-arm navigated modified Goel procedure with postoperative halo-vest immobilization. At one-year follow-up, good neurological recovery and solid bone fusion were observed. The patient had no complications such as epidural hematoma, infection, or nerve or vessel injury. This novel procedure is a useful and safe technique that protects surgeons and staff from radiation risk.

Key words: atlantoaxial fixation, Down syndrome, C-arm free, O-arm navigation surgery, modified Goel technique

Down syndrome (DS) is a genetic disorder caused by trisomy of the 21st chromosome, and is the most common of all genetic malformation syndromes [1]. The symptoms of DS are characteristic facial features, hypotonia, intellectual disability, cardiorespiratory diseases, and digestive-system problems [2]. Atlantoaxial subluxation (AAS) refers to a loss of stability and compression of the spinal cord between the C1 posterior arch and odontoid process, which can cause cervical myelopathy. AAS occurs in 10-30% of DS patients, but only 1-2% of patients with AAS with DS are symptomatic [3,4]. Asymptomatic AAS can be treated with a cervical collar or observation. However, conservative treatments for symptomatic AAS are ineffective, and so surgery is indicated for such patients [5]. There is no uniformly accepted method for determining treatment strategies [6]. Some studies recommend surgery even for asymptomatic AAS to prevent development of myelopathy, due to concerns about increased morbidity and mortality [7]. The main procedures reported in the literature are posterior atlantoaxial fusion (PAF) with various techniques [8,9].

There are several problems related to conventional PAF in DS, including radiation exposure and complications such as vertebral artery injury (VAI) and nonunion [6]. The use of fluoroscopy is necessary for screw insertion. Radiation to surgical staff is currently con-

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sidered a serious problem, and several reports have sought to address it by developing C-arm free techniques for spinal surgeries [10,11]. Here, we report a novel technique of C-arm free O-arm navigated PAF with O-arm navigation guidance.

Case Report

Institutional ethical committee approval was obtained for this study (No 288). A 5-year-old boy with DS was referred to our orthopedic department in May 2018 with progressively worsening neck pain and gait disturbance. At 1 year and 9 months of age, he was diagnosed with AAS, and was counseled to avoid strenuous exercise. AAS worsened, and he would cry with pain while washing his hair with his neck flexed. In the examination, neck pain prevented him from flexing his neck. While he had unsteady gait and imbalance, there was no muscle weakness of the limbs. Hyperreflexia of the bilateral arms and legs was observed, but Babinski reflex was negative.

Preoperative radiograms showed AAS with an atlantodental interval (ADI) of 10 mm, and the subluxation was irreducible with extension (Fig. 1). Preoperative computer tomography (CT) revealed Os odontoideum and atlas rotatory subluxation (Fig. 2A-C). Preoperative magnetic resonance imaging (MRI) indicated spinal cord compression between the C1 posterior arch and odontoid process (Fig. 2D).

Operative Procedure

Patient positioning and exposure. The patient was positioned prone on a Jackson frame with a full carbon skull clamp to enable the O-arm scan. The procedure was performed under neuromonitoring. The atlas and axis were exposed with a 5-cm posterior midline incision. The reference frame for the navigation was fixed at the C2 spinous process. The O-arm was then positioned, and three-dimensional (3-D) reconstructed images were obtained, transmitted to the StealthStation navigation system Spine 7^{R} (Medtronic Sofamor Danek, Minneapolis, MN, USA) and integrated.

Modified Goel technique. After verifying every navigated spinal instrument, the accuracy of navigation should be checked. The entry point for the C2 pedicle screw was marked, and a high-speed burr was used to make the hole without pushing the C2 and further compressing the spinal cord. A navigated pedicle probe and tap were used, and the C2 pedicle screw was inserted. The entry point for the C1 lateral mass (LM) was marked with the navigated probe (Figs. 3,4). A gutter was made in the C1 posterior arch and a hole was made in the C1 LM with the navigated burr and drill. Then, the C1 LM screw was inserted using Lee's notching technique (Fig. 5) [12].

Intraoperatively, other O-arm images were obtained to check the correct placement of screws. Rods were

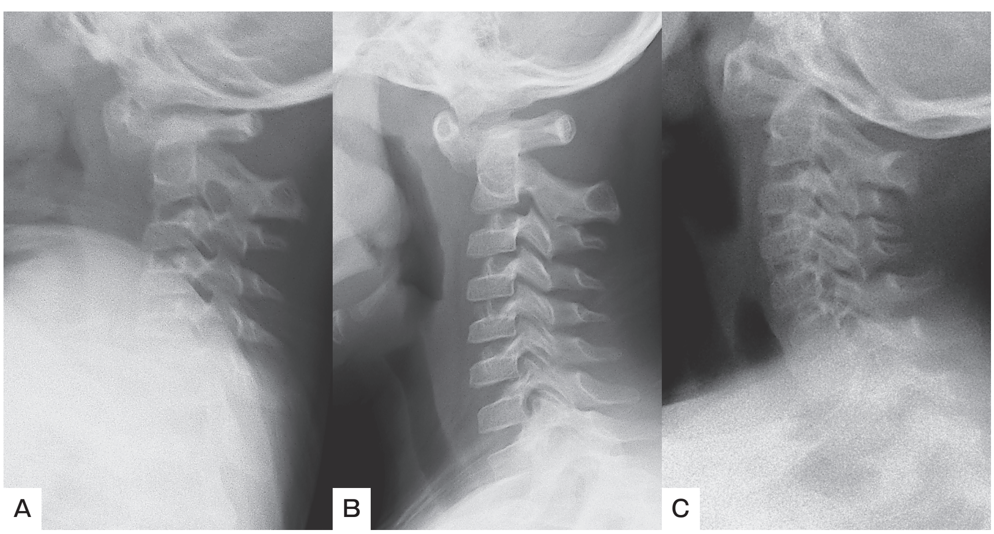

Fig. 1 Preoperative radiograms. A, Cervical lateral neutral radiograph; B, Cervical lateral flexion radiograph; C, Cervical lateral extension radiograph.

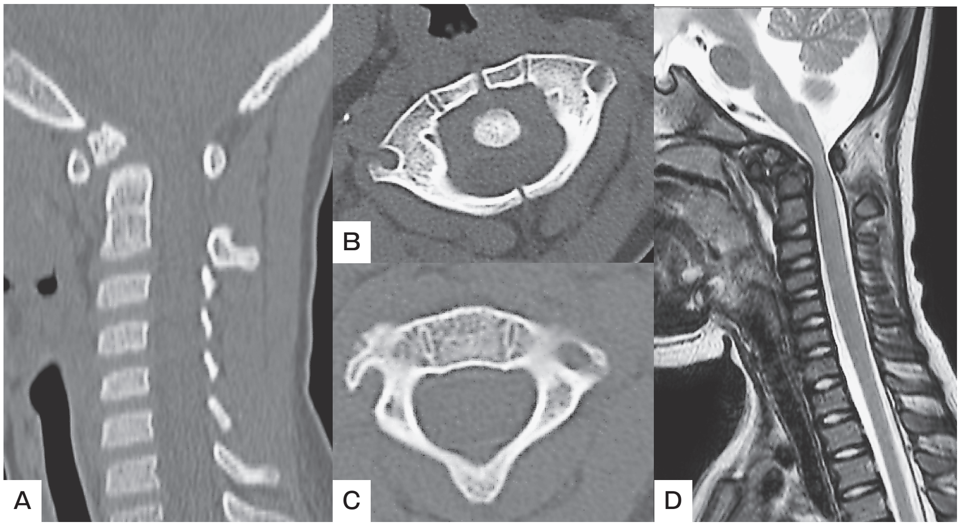

Fig. 2 Preoperative CT and MRI. A, Sagittal reconstruction CT; the Os odontoidium was identified; B, Axial CT at the C1 level showing rotatory subluxation; C, Axial CT at the C2 level; D, T2 weighted mid-sagittal MRI. An MRI mid-sagittal cut showing spinal cord compression at the C1 level.

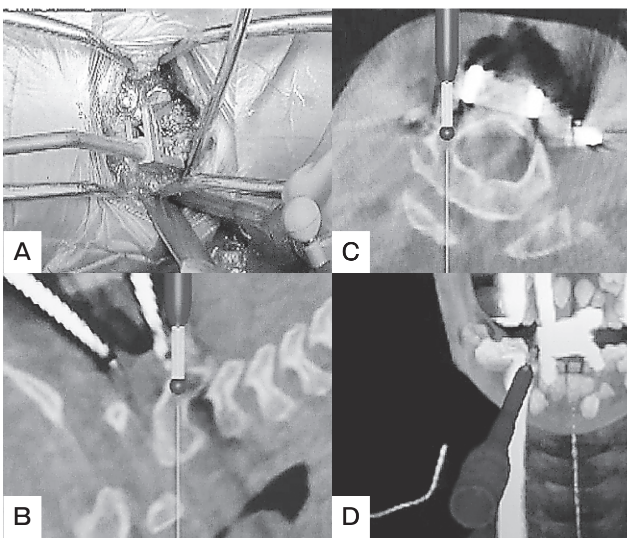

Fig. 3 C2 pedicle screw preparation with a high-speed burr. A, Intraoperative image; B, Sagittal navigational image; C, Axial navigational image; D, 3-D navigational image.

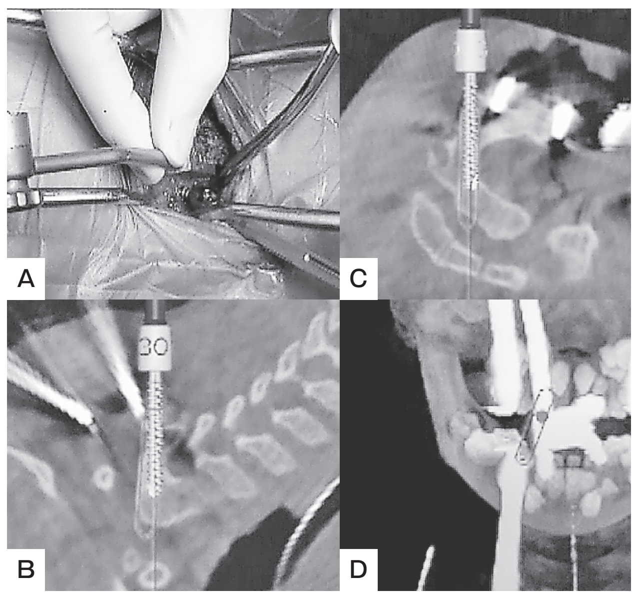

Fig. 4 C2 pedicle screw. A, Intraoperative image; B, Sagittal navigational image; C, Axial navigational image; D, 3-D navigational image.

attached and subluxation was reduced with an implant maneuver. Care was taken to make sure not to inadvertently increase the gap at C1/2 when the screws were tightened to the rods (Fig. 6). An iliac crest bone graft was inserted and a halo vest applied to immobilize the neck (Fig. 7).

Post-operative results. Operative time was 2 h 13 min, and estimated blood loss was 50 ml, with no intra- or postoperative complications. Two days post-surgery, the patient could walk independently, and by two months he was back to full activity. Postoperative radiographs showed the screws were in the intended position, securing the cortex of the anterior arch of C1 and maintaining an appropriate sagittal spine alignment (Fig. 8). At two months follow-up, the patient was asymptomatic, and a follow-up CT scan

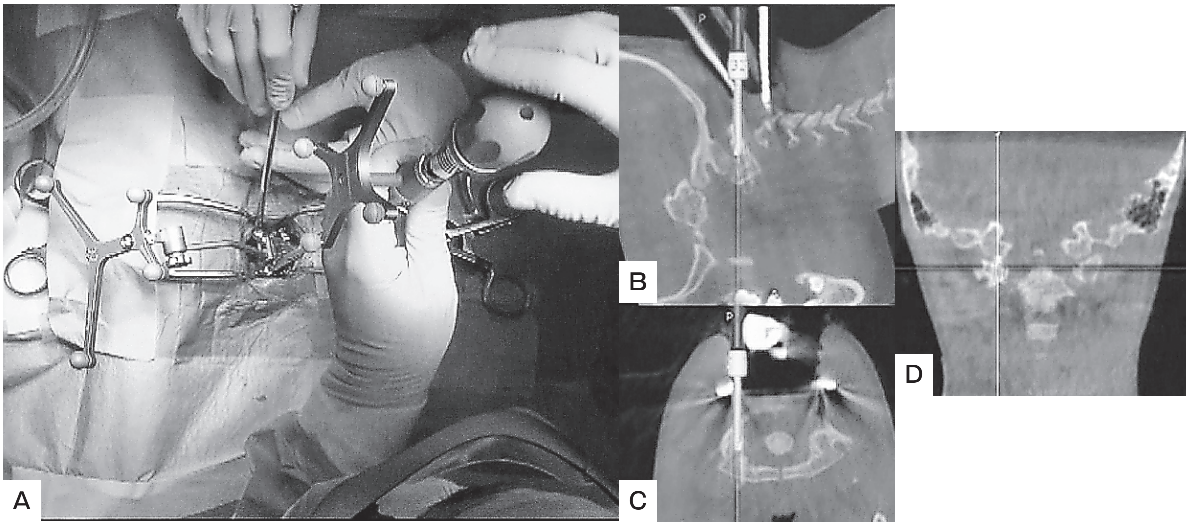

Fig. 5 C1 lateral mass screw. **A**, Intraoperative image; **B**, Sagittal navigational image; **C**, Axial navigational image; **D**, Coronal navigational image.

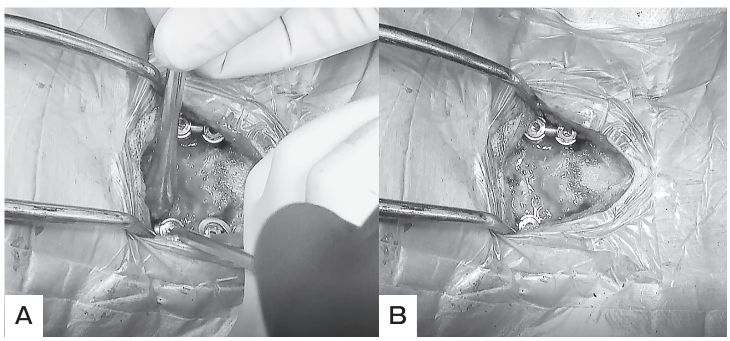

Fig. 6 Rod insertion and reduction. **A**, Reduction with a persuader; **B**, After rod maneuver reduction.

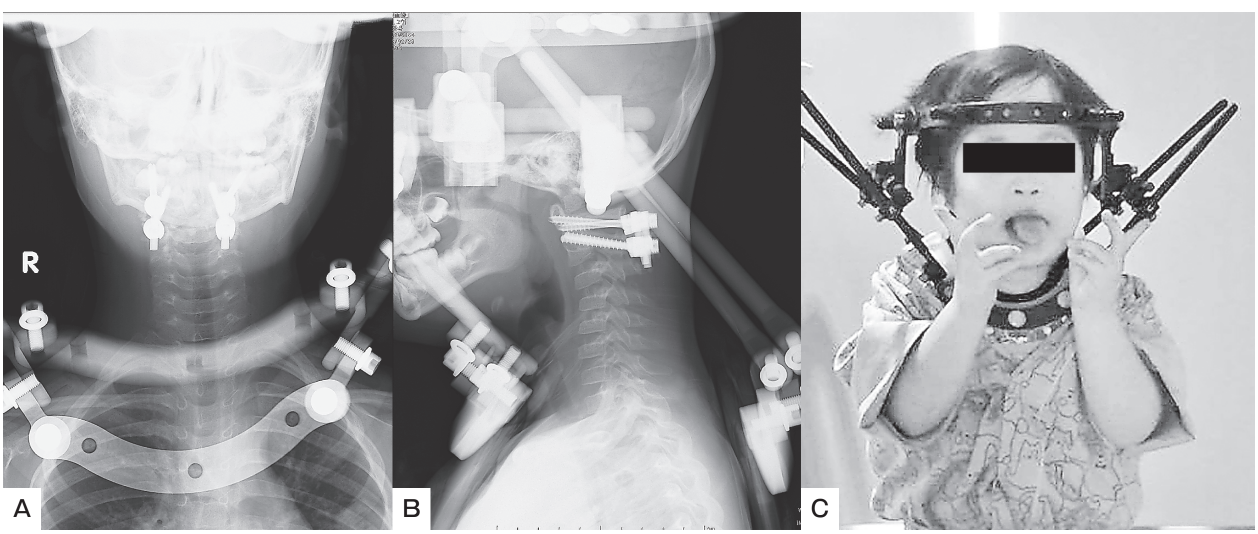

Fig. 7 Postoperative images with Halo-jacket. **A**, Anteroposterior radiogram; **B**, Lateral radiogram; **C**, Halo-jacket.

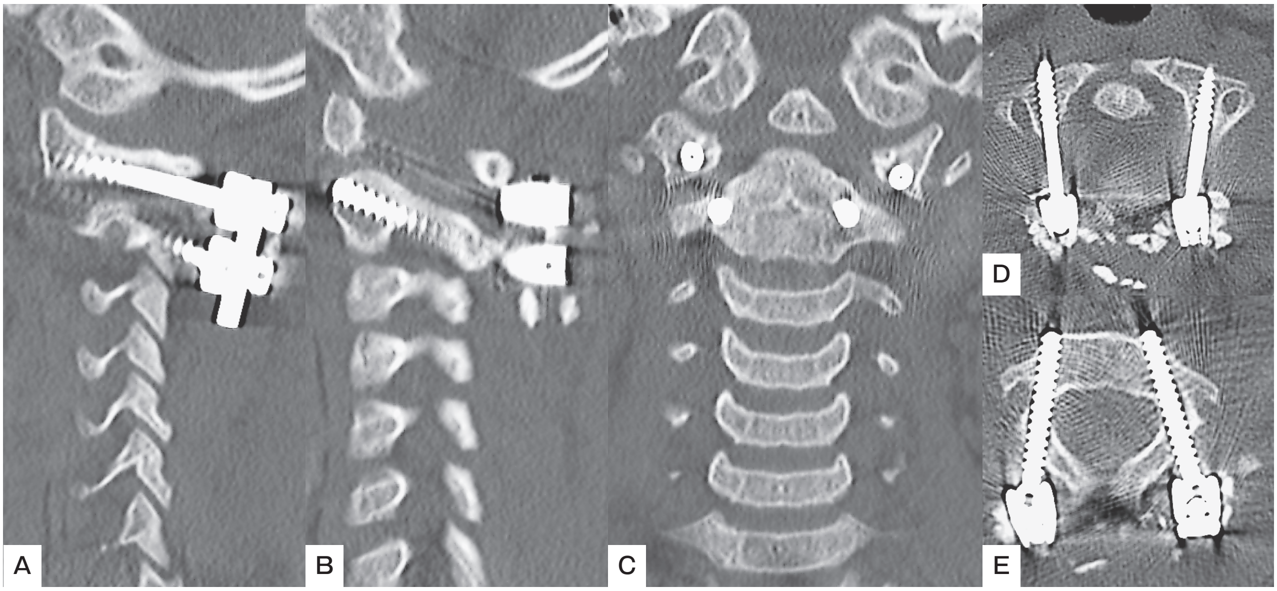

Fig. 8 Postoperative CT. A, Right parasagittal reconstruction CT; B, Left parasagittal reconstruction CT; C, Coronal parasagittal reconstruction CT; D, Axial view at the C1 level; E, Axial view at the C2 level.

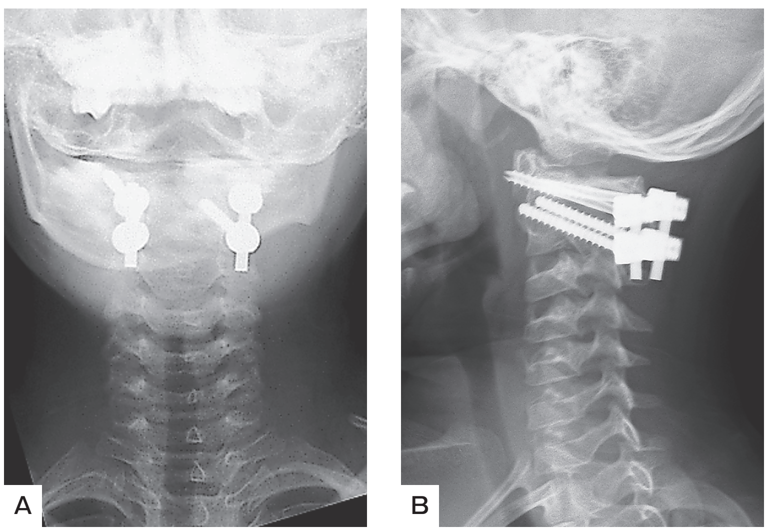

Fig. 9 Final follow-up radiograms. A, Anteroposterior view; B, Lateral view.

revealed no implant loosening. At one-year follow-up, the patient was symptom free with CT scan and radiograms showing solid fusion (Fig. 9).

Discussion

DS is the most common genetic cause of intellectual disability [1]. It occurs in approximately 1 in 700-900 live births, and incidence increases with maternal age [13]. Children with DS may have associated defects of the heart, respiratory system, and thyroid, and 10-30% may have AAS, although only 1-2% of these cases will be symptomatic [3,4]. While asymptomatic patients can be simply observed, surgical treatment is mandatory for symptomatic cases. The clinical course of asymptomatic cases is unknown, and some studies have advocated surgery even for these cases to prevent the development of myelopathy [7]. Historically, sublaminar wiring with bone grafting was tried, but fusion rates were low [8]. In DS, the fusion rate is further reduced due to the hypermobility of patients and inherent ligamentous defects [5].

Various techniques have been applied for PAF in DS [6,8]. Wright's technique (lateral mass screws and laminar screws) is relatively safe but biomechanically less stable [14]. The Magerl technique is reported to produce the strongest biomechanical fixation, but reduction should be obtained before inserting the screws [15]. With this technique the rate of VAI was reported to be as high as 4.1% due to drill penetration through the lateral joint [16]. Goel reported PAF with plates, C1 LM screw, and C2 pedicle screw fixation [8]; Harms then modified this technique with 2 rods [9].

The modified Goel technique has several advantages. First, the reduction can be achieved after placement of 4 screws with strong reduction force. Second, there is less risk of VAI because screws are inside the cancellous bone and there is no need to penetrate the cortex. The VAI rate of the modified Goel technique has been reported as 0.34% [17]. The C1 LM screw also involves a risk of vascular injury (0.26%), and the rate of unacceptable position of the C1 lateral mass screw was reported to be 2.8% [18]. Tan, Goel and Lee described different techniques for C1 lateral mass screw fixation with slight variation in entry point (Fig. 10) [8,12,19]. With the Tan and Goel techniques, the risk of VAI and C2 nerve injury was higher. Therefore, the notching technique by Lee is preferred [12].

Previous studies have shown that very high rates of fusion, from 92% to 100%, can be achieved in non-DS pediatric patients undergoing PAF [21,22]. The risk factors of C1/2 nonunion were etiology of subluxation,

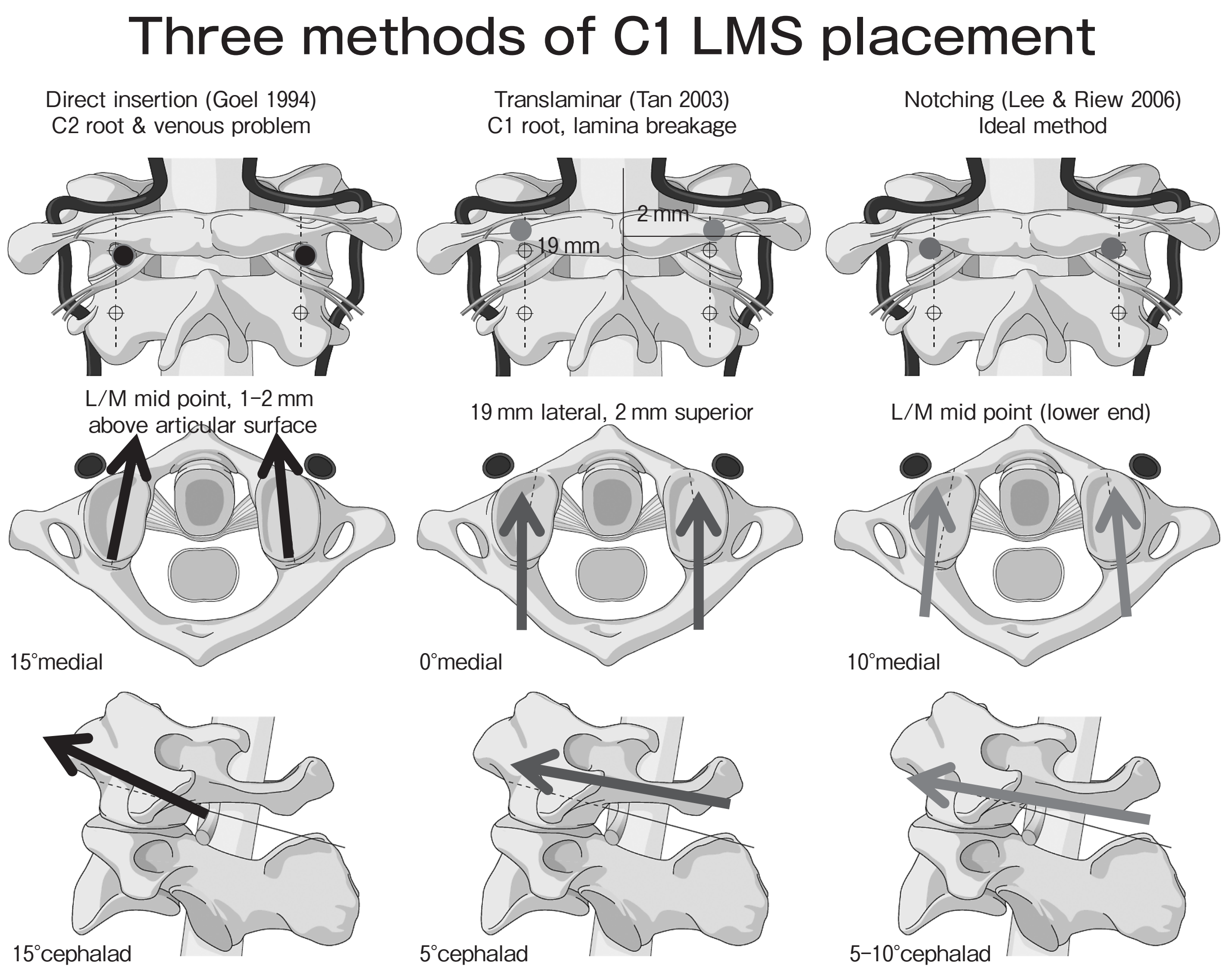

Fig. 10 Three methods of C1 lateral mass screw placement.

unilateral fixation, and no brace after surgery. In a multicenter cohort study of 131 pediatric patients, Brockmeyer reported that DS was a significant risk factor predictive of surgical nonunion (47%, odds ratio 14.6) [22]. To prevent nonunion with PAF in DS, several approaches have been tried. The halo vest is a good external immobilization device and is widely used for older children and adults [23]. However, it is very difficult to expect an active young DS patient to cooperate and move carefully under the restriction of the halo vest. Tseng reported successful use of halo-vest immobilization for a DS child (32-months-old) for 3 months [24]. In our case, the patient was immobilized with the halo vest for two months, followed by a Philadelphia collar for another two months.

One problem with the modified Goel technique is that it requires fluoroscopy to position the screws. With conventional methods, the trajectory and length of the C1 LM screws should be confirmed by fluoroscopy, because C1 is very mobile and the VA is located very near the C1 posterior arch. With our new technique, the reference frame is attached to the C2 spinous process, and the entry and hole for LM screws are gently made in the LMs with a navigated high-speed burr. Thus, C1 is less likely to move and the C1 posterior arch prevents injury of the V3 and V4 parts with Lee's notching technique.

It has been reported that malignancies are increasing in orthopedic surgeons due to radiation [25]. Our procedure is C-arm free O-arm navigated, and therefore eliminates radiation exposure of the operating surgeon and staff. There might be concerns about patient radiation exposure with O-arm use. It is estimated that one O-arm scan delivers 9 mGy of radiation, which is equivalent to 35 sec of fluoroscopy [26]. One screw placement under fluoroscopy takes 7-20 sec [27]. Thus, overall exposure to a patient receiving four screws with O-arm navigation is equivalent to that with fluoroscopy.

There are some limitations to this procedure. The accuracy of navigation can be hampered if the reference frame moves. Repeated O-arm scans might be needed in such situations. Another potential problem is the penetration of halo vest pins; specially designed pins are necessary for a pediatric population.

Conclusion

This novel procedure is a useful and safe technique. It allows for correct placement of screws under navigation guidance without exposing surgeons and staff to the risk of radiation.

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