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Basilar Invagination and Atlantoaxial Dislocation: Reduction, Deformity Correction and Realignment Using the DCER (Distraction, Compression, Extension, and Reduction) Technique With Customized Instrumentation and Implants

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Objective: The technique of distraction, compression, extension, and reduction (DCER) is effective to reduce, realign, and relieve cranio-spinal compression through posterior only approach.

Methods: Study included all patients with atlantoaxial dislocation and basilar invagination (BI) with occipitalized C1 arch. Study techniques included Nurick grading, computed tomography scan to study atlanto-dental interval, BI, hyper-lordosis, and neck tilt. Sagittal inclination (SI), coronal inclination (CI), cranio-cervical tilt, presence of pseudo-joints, and anomalous vertebral artery were also noted. Patients underwent DCER with/without joint remodeling or extra-articular distraction (EAD) based on the SI being $< 100^\circ$, 100° – 160° , or $> 160^\circ$ respectively. In cases with pseudo-joints, joint remodeling was performed in type I and EAD in type II. Customized 'bullet shaped' PSC spacers ($n = 124$) and prototype of the universal craniovertebral junction reducer (UCVJR, $n = 36$) were useful.

Results: A total of 148 patients with average age 27.25 ± 17.43 years, ranging from 3 to 71 years (87 males) were operated. Nurick's grading improved from 3.14 ± 1.872 to 1.22 ± 1.17 ($p < 0.0001$). Fifty-two percent of total joints ($n = 154/296$ joints) were either type I (19%)/type II (33%) pseudo-joints. All traditional indices such as Chamberlain line, McRae line, atlanto-dental interval, and Ranawat line improved ($p < \text{at least } 0.001$). BI, SI, and CI values correlated with type of pseudo-joints ($p < 0.0001$). Side of neck tilt correlated with the type of pseudo-joint ($p < 0.0001$). Cervical hyperlordosis improved significantly ($p < 0.0001$).

Conclusion: Occipito-C2 pseudo-joints are important in determining the severity of BI. Asymmetrical pseudo-joint causes coronal/neck tilt. Type of pseudo-joint can strategize by DCER. Customized instruments and implants make technique safe, effective and easier.

Keywords: Basilar invagination, Atlantoaxial dislocation, Vertebral artery, Distraction, Compression, Extension



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INTRODUCTION

DCER stands for distraction, compression, extension, and reduction. This is a surgical technique to reduce, realign and

correct (even very severe) basilar invagination (BI), atlantoaxial dislocation (AAD) with a posterior only, single staged approach. This involves motion in 2-axis using the lever principle. This is a technique which was pioneered by the senior author (PSC).¹⁻⁴

The craniovertebral junction (CVJ) is composed of osseous complex allows significant mobility while maintaining biomechanical stability.⁵ Developmental CVJ anomaly usually consists of AAD and BI associated with the occipital fusion of C1 arch. It is a complex pathology if untreated leads to disabling neurological deficits.

BI and AAD (occurring in the setting of BI) usually do not reduce on skeletal traction. The current philosophy is to reduce the deformity, realign, and relieve the spinal cord decompression intraoperatively from a single posterior approach only.⁶⁻⁸

It is important to understand that CVJ anomalies occur in roughly in 2 situations.

- (1) The C1 arch is not fused with the occiput: Such situation usually produces AAD, which may be reducible. The treatment of choice is C1 lateral mass and C2 pars screw fixation as described by Goel and Shah.⁹ Such patients usually do not have BI unless an underlying pathology like osteoarthritis destroys and reduces the height of facets leading to the invagination of the dens upwards. A bone graft is generally recommended as a spacer unless the patient also has BI in which case, a metal/PEEK spacer may be advised.
- (2) C1 arch is fused with occiput: Such cases usually present with moderate to severe BI. Such cases are also associated with a significant increase in the incidence of vertebral artery (VA) anomalies.¹⁰ It is in such cases that the technique of DCER is very useful not only to reduce the AAD and BI but also to correct the hyper-lordosis of the subaxial cervical spine. The author will also describe certain unique instrumentation developed by him to allow the surgeon perform this technique more easily and effectively.

MATERIALS AND METHODS

1. Indications of DCER

DCER is currently indicated in conditions with BI and AAD in developmental anomalies with a fusion of C1 arch.

2. Contraindications

These may be divided into absolute (not possible to overcome), relative (difficult to overcome).

1) Absolute contraindications (not possible to overcome)

These include:

- (1) Clival segmentation anomalies: Here, the entire clivus forms a continuous bony mass or has a pseudo-articula-

tion (Fig. 1A-C) and is associated with a ventral compression of the cord.

- (2) Severe BI associated with platybasia sans AAD: In such conditions the entire odontoid process telescopes inside along with clivus. Obviously, a transoral/lateral nasal approach will be required even though such a surgery would be very challenging. Fortunately, such anomalies are rare (Fig. 1D-F).
- (3) Infective pathologies^{11,12}: Conditions like tuberculosis (Fig. 1G) are associated with large-scale bony destruction. These, hence require both ventral and dorsal approaches.

2) Relative contraindications

These contraindications are related to the expertise and experience of the surgeon in performing DCER. These include:

- (1) Presence of vertical joints: Earlier, this was an absolute contraindication to posterior only approach. However, with description of extra-articular distraction with DCER by the author this type of pathology may be optimally treated through a posterior only approach. However, this should be performed only by surgeons with significant expertise in technique of DCER.
- (2) Presence of VA over the occipito-C1/C2 (OC1-2) complex: All cases of bony CVJ anomalies should undergo computed tomography (CT) angiogram with 3-dimensional (3D) reconstruction to ascertain the position of VA (Fig. 1H). In the presence of anomalous course of VA being present, especially directly over the joint complex, mobilization of VA may be performed.

3. Investigations

Apart from magnetic resonance imaging (MRI), it is mandatory to perform CT-scan with sagittal and coronal reconstruction (thin-slice CT scans: 0.63 mm or 0.7 mm). In addition, a CT angiogram with 3D reconstruction is also mandatory. CT angiogram provides the information of (1) lie of VA and (2) side of dominance of VA. The nondominant side should be instrumented first.

Bone quality is assessed by good quality plain X-rays. In case of doubt, a bone densitometry may be performed. In cases of severe osteopenia or osteoporosis, it may be worthwhile to wait while providing medical therapy to enhance the strength of the bones. Another useful strategy is to enhance the strength of neck muscles from outpatient department itself (isometric neck strengthening exercises).

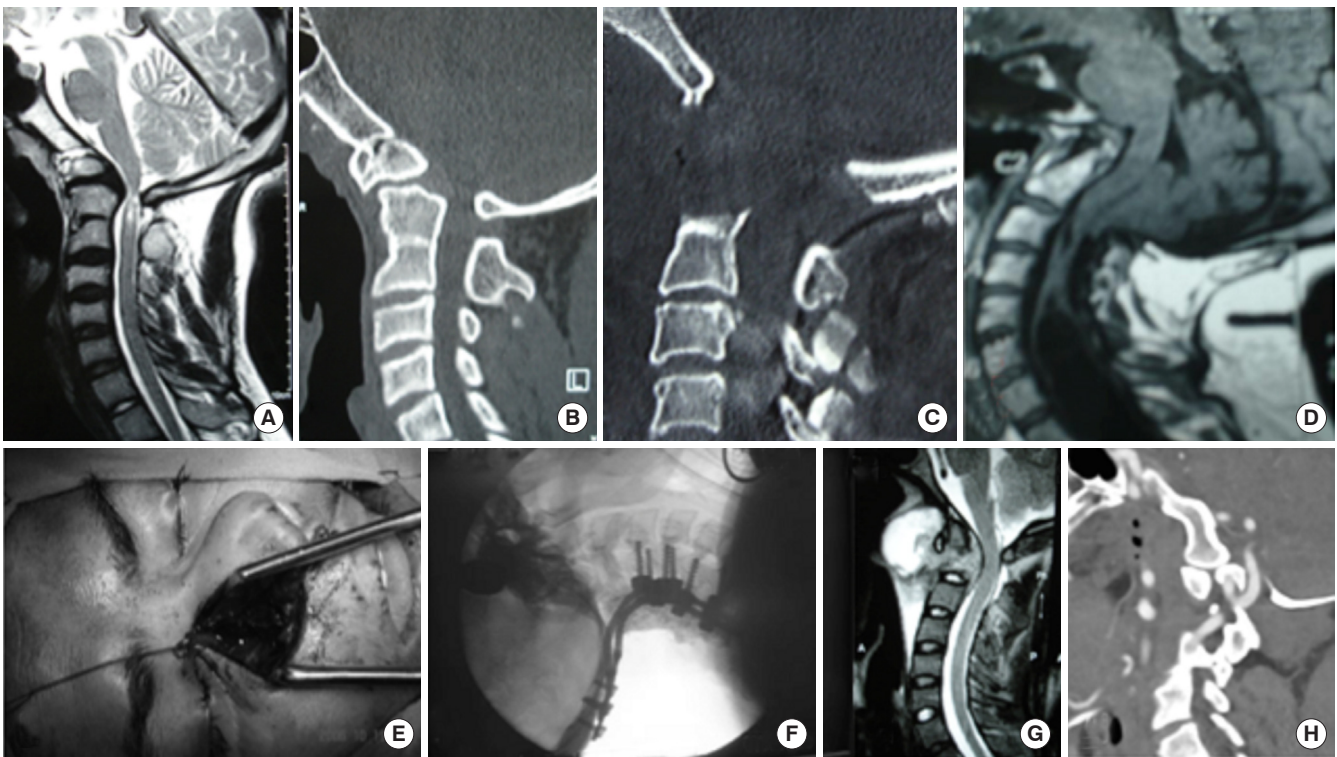


Fig. 1. The following figures show the absolute indications for a transoral surgery in the current scenario. (A) Clival segmentation anomaly with ventral compression in magnetic resonance imaging and (B) computed tomography scan. Panel C shows the same case following a transoral excision of odontoid surgery with posterior fixation. Panel D shows severe basilar invagination with platybasia without atlantoaxial dislocation. (E, F) Note the severe Chiari with massive syrinx. (G) The same case underwent a transnasal approach, excision of dens and long segment posterior fixation. (G) Infective pathologies with large abscess ventrally as in tuberculosis is another indication of a transoral procedure. Presence of vertebral artery (VA) over the joint line (H) as shown here is a relative contra-indication. This may be overcome by mobilizing the VA and then performing DCER (distraction, compression, extension, and reduction).

4. Surgical Planning

It is important to plan the surgical procedure based on all investigations (MRI, CT scan with angio and good quality digital plain X-rays).

5. Measurement of All Indices

It is important to measure all the indices prior to surgery in all patients. These include the traditional indices & special indices (as described by the author). The indices are important: (1) to measure the deformity and cord compression before and after surgery, (2) to measure the degree of deformity which is to be corrected (see later).

1) Traditional indices

- (1) Chamberlain line: dens being about 2.3 ± 2.6 mm below this Line.¹³⁻¹⁶
- (2) Wackenheim's line¹⁴: being about 0.9 ± 2.2 mm below this line.

(3) McRae line¹⁵: being about 5.8 ± 1.6 mm below this line.

(4) Modified Ranawat line^{15,16}: from the midpoint of the base of C2, a line was drawn to meet the line drawn from the center of the anterior arch of C1 to the center of the posterior arch (normal value: 29.7 ± 2.6 mm; < indicates BI).

(5) Clivus canal angle^{15,16}: The realignment was measured using the clivus canal angle (normal value $> 150^\circ$).

2) Special indices

These indices have been described by author and relate to the morphology of the joint.^{1-4,17-19} Here, the sagittal inclination (SI) and craniocervical tilt (CCT) correlated significantly with the severity of the BI and AAD ($p < 0.001$). Coronal joint inclination (CI) was also significant ($p = 0.003$) but was less than SI and CCT. It correlated with BI only.

(1) Sagittal joint inclination (Fig. 2A-C): The normal value is $87.15^\circ + 5.65^\circ$ (approximately 90° , i.e., the joint line is parallel to the ground). It increases significantly with increas-

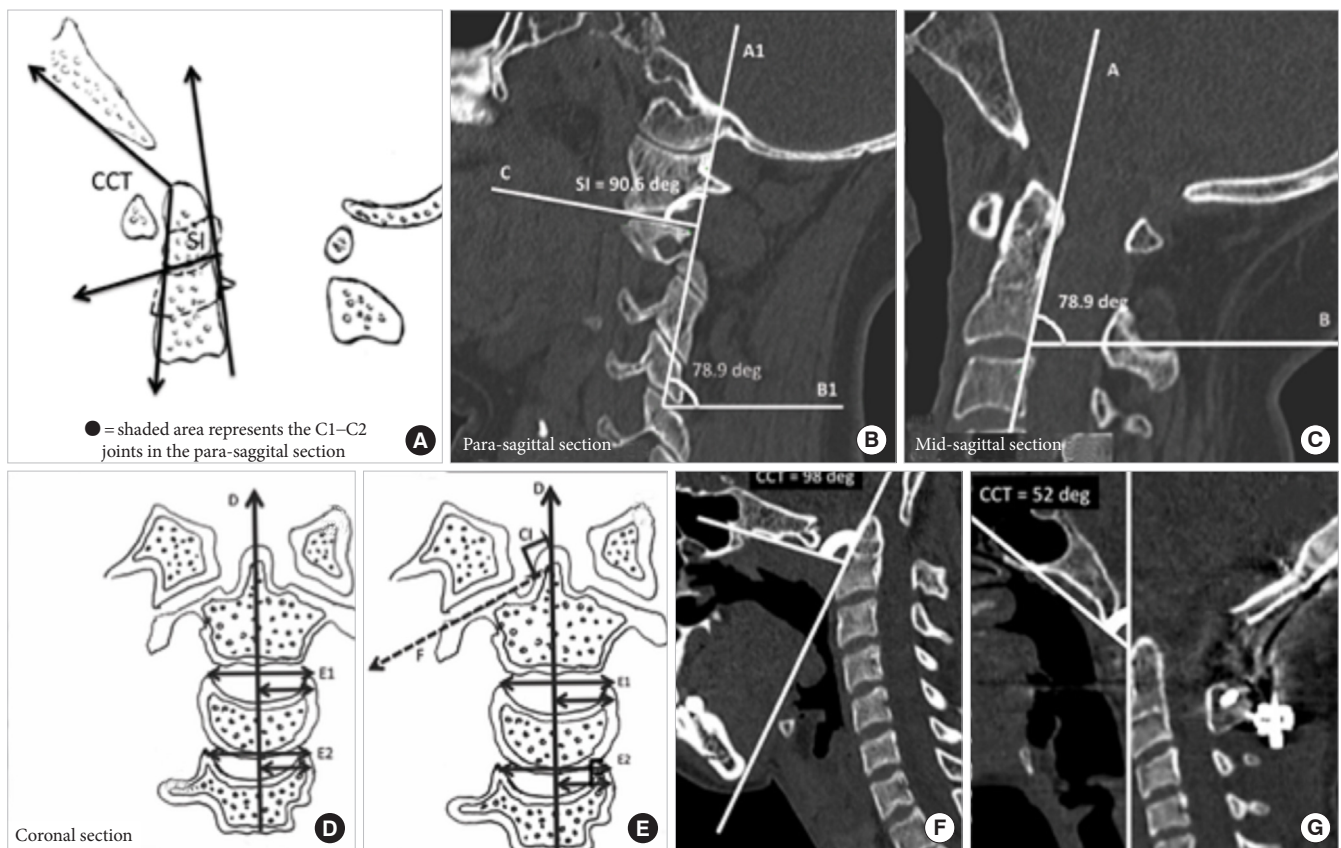


Fig. 2. Panel A shows the 2 important indices which correlate with degree of severity of basilar invagination (BI) and atlantoaxial dislocation (AAD), these being sagittal inclination (SI) and BI. Panels B and C show the method to calculate SI: SI is actually the angle between the long axis of odontoid process and the surface of the C1–2 joint. However, since the odontoid process is in the midline and the C1–2 joint is at the level of parasagittal section, the following figure shows the method to measure this angle. Step I (C): In the mid sagittal section, a line is drawn along the posterior border of the odontoid process (line A). Next, a line is drawn parallel to the border of the image (line B), which now subtends an angle “ang” (C). The value of “ang” is 78.9° in panel C. Step II (B): This is performed in the para sagittal section where the joints are seen. In this step, a line is first drawn parallel to the border of the image (line B1, B). Now the same value of angle is constructed as in “ang” (called here as “ang1”, C) with another line A1 that now passes along a point on the posterior border of the C2 joint surface. Step III: This is also performed in the para sagittal section (B). In this step, a line is now drawn passing parallel to the C2 facet joint (line C, B). The angle now subtended between the lines A1 and C is called as the SI. In this case, the SI is about 90.6°. In our series, values of SI in normal individuals were $87.15^\circ \pm 5.65^\circ$; in patients with BI and AAD were $127.1^\circ \pm 22.05^\circ$ ($p < 0.01$ for both BI and AAD). Please note: The horizontal line B that is drawn in the mid sagittal section (B, C) should be parallel to the border of the film. It may be drawn at any distance from the lower border of the image on the film. Since the entire computed tomography is performed on the patient at the same time point of time, this line will have the same referential value in mid sagittal and para sagittal sections. Hence this line is useful to measure the angle between the long axis of odontoid process and the C1–2 joint, which is the SI. (E) This figure shows the method to measure the coronal joint inclination, which is actually the angle between the long axis of odontoid process and the C1–2 joints in the coronal plane. In the midcoronal section, where the C2/C1 joint is seen well, midpoints of 2 lines joining the uncinate processes of C2 and C3 respectively are first marked (here being E1, E2). A line joining these 2 points (called line D) is now drawn extending upwards. Another line (line F) is now drawn parallel to the upper border of the C2 joint, which now joins the line D. The angle subtended between the line D and F is called the coronal joint inclination. The normal value of CI in our series was $110.3^\circ \pm 4.23^\circ$ and in patients with BI and AAD was $121.15^\circ \pm 14.6^\circ$ (mean p-value between right and left joints was 0.2). Finally, panels F and G show the improvement of craniocervical tilt (CCT) following DCER (distraction, compression, extension, and reduction). Also to be noted is the improvement in curvature of the subaxial spine. Reprinted from Chandra, et al. *Oper Neurosurg* 2014;10:621-30.¹

ing severity of BI and AAD ($p < 0.001$). The Fig. 2A-C show how to measure the SI.^{1,17,18}

- (2) Coronal joint inclination (Fig. 2D, E): The normal value has been assessed as $110.3^\circ \pm 4.23^\circ$. It correlates only with BI ($p = 0.03$). This parameter to be useful correct the coronal tilt. However, it has been found to be of less significance.^{1,2,17,18}
- (3) Craniocervical tilt (Fig. 2A, F, G): This is a measure of severity of sagittal deformity (Fig. 2F) and shows the degree of fixed deformity (normal: $60.2^\circ \pm 9.2^\circ$; is increased in BI usually $> 80^\circ$) The technique of measurement is shown in Fig. 2A. As shown in Fig. 2F, G, it may be seen that this deformity improves significantly following DCER. Also, it to be noted is the significant degree of correction of the curvature of subaxial spine.^{1,2,17,18}

6. Planning Surgical Strategy; Types of DCER

Based on degree of joint inclination (SI), it is next important to assess the type of surgery^{3,4} (Fig. 3A-C) to be performed (see later for technique of surgery).

1) Type I surgery

In cases, where the SI ranges from $90^\circ - 100^\circ$ (i.e., the joint line is more or less parallel to the ground in standing position), a

DCER only would suffice (Fig. 3A). This fundamentally means exposure of the joint surfaces, drilling the surfaces using a diamond drill to denude the cartilage (the author prefers to call it 'optimization' of joint surface) and placement of the C1-2 spacer. The compression and extension also required is not much and a short segment fixation between occiput and C2 translaminar screw will suffice. However, more recently, the author prefers to a 3-point fixation, i.e., an implant between occiput, C2/3 and/or C3/4. C3 and C4 fixation is performed if for some reason, a C2 pars screw cannot be placed. In case of a 2-point fixation, the author prefers a C2 translaminar screw as it provides a greater lever length. In cases, if the surgeon wants to perform a short segment fixation (occipito-C2), the author will prefer a C2 trans-laminar screw as this would have lesser screw-bone interface tension due to a longer lever length (unlike a C2 pars screw which places a much higher stress on the screw bone interface due to a shorter lever arm length; see later).

2) Type II surgery

In cases, where the SI ranges from $100^\circ - 160^\circ$ (i.e., becomes more angulated but falls short of becoming vertical), a joint remodeling (Fig. 3B) should be performed, followed by DCER (see Supplementary video clip 1). Here the posterior lip of the joint surfaces is drilled using a diamond drill so as to make the

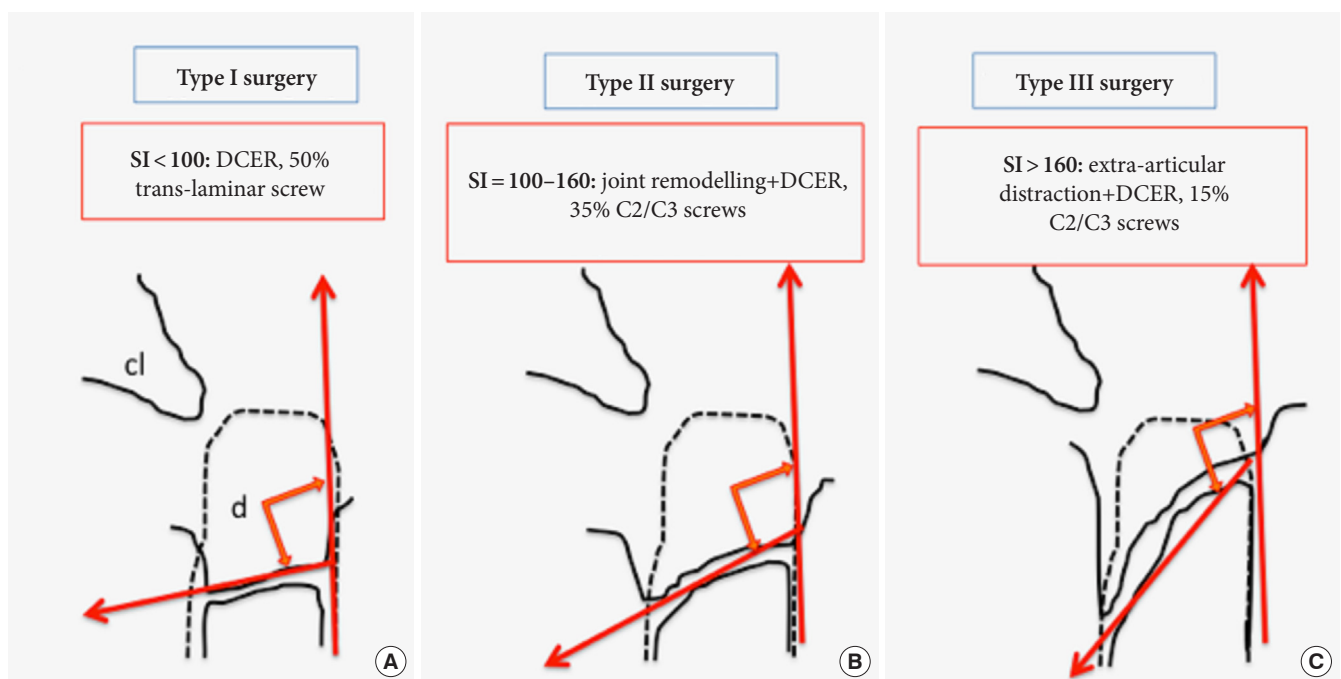


Fig. 3. Schematic diagram showing different types of surgeries depending on the sagittal inclination (SI). d, dens; cl, clivus; DCER, distraction, compression, extension, and reduction.

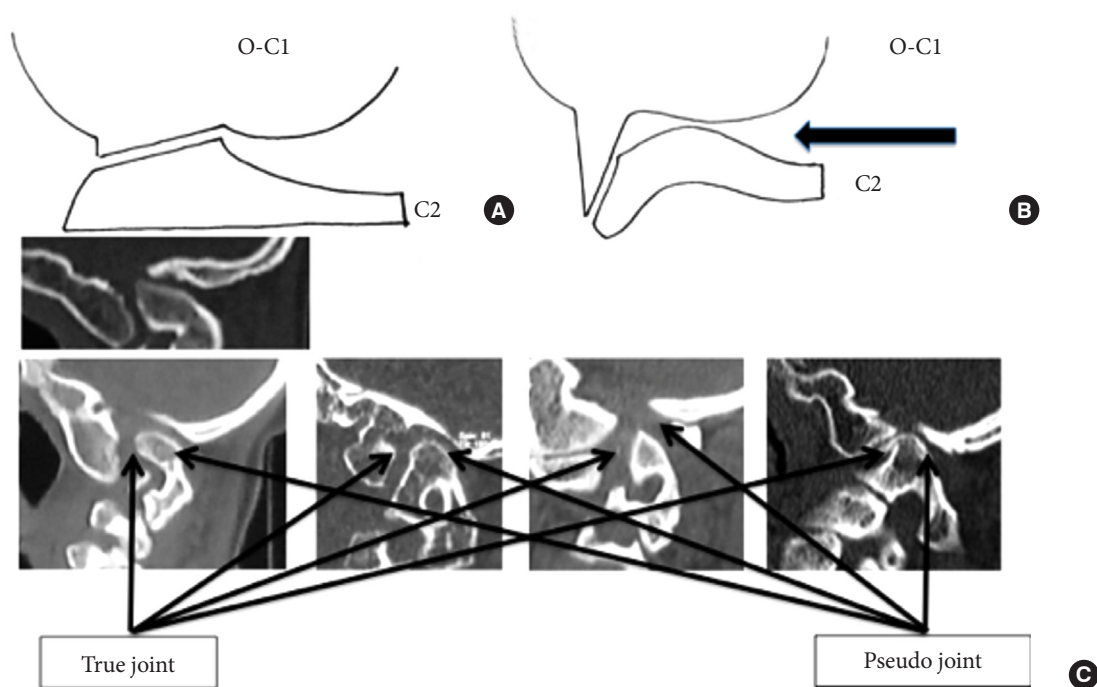


Fig. 4. Schematic diagram showing the concept of formation of pseudo-joint. Panel A shows the normal configuration with normal sagittal inclination (SI). Once the SI becomes $>160^\circ$, the occiput sinks, thus coming in touch with superior surface of the C2 pars forming a pseudo-joint (B). Once this is understood, it may be seen that this occurrence is an anatomical surety. Panel C shows various examples of pseudo and true joints.

joint surfaces parallel to each other and also to convert the value of SI back to around 90° .

3) Type III surgery

Here the SI ranges between 160° – 180° , i.e., the joint is almost vertical/ vertical. Here, an extra-articular spacer is placed within the 'pseudo-joint' to perform distraction. This is then followed by technique of compression and extension (extra-articular distraction with DCER). The concept of spacer placement within the pseudo-joint is again a first of its kind, described by the author. It is important to identify the pseudo-joint properly (see next).

Currently, the author in almost all cases, 'optimizes' the joint surfaces by drilling the surfaces with a diamond drill so that the placement of the spacer can be made more effective. The coronal inclination (CI) is particularly effective to plan the coronal correction. In a vast majority of cases, the CI is unequal on both sides indicating a coronal tilt to one side. Correction of the deformity would involve placement of different height of spacers on either side.

4) Identification of pseudo-joint

In cases, where the SI is more than 160° , it is now obvious

that the vertical joint is incapable of transmitting the weight of the head. To compensate this, in such cases, invariably there is formation of a 'pseudo-joint' between the occiput and the superior surface and proximal part of the C2 pars (Fig. 4). It is important to identify the position of the pseudo-joint, which may be used for placement of spacer to perform the DCER technique. Since the pseudo-joint is placed posterior to the true joint, it is much easier to identify and place spacer within the pseudo-joint provided the surgeon can understand the anatomy properly. The author again, always optimizes the joint surfaces using a diamond drill before placement of the spacer.

5) Types of pseudo-joints

The author with his experience has again identified 2 types of pseudo-joints. These include type I pseudo-joint: These had an oblique orientation of C1–2 joint and partial contact between occipito-C2, type II pseudo-joint: these had near-vertical/complete vertical orientation of C1–2 joint leading to weight transmission through pseudo-joint occipito-C2 joint (Fig. 5).

6) Position of VA

Assessing the position of VA is very important. This is performed with 3D CT scan with coronal and sagittal reconstruct-

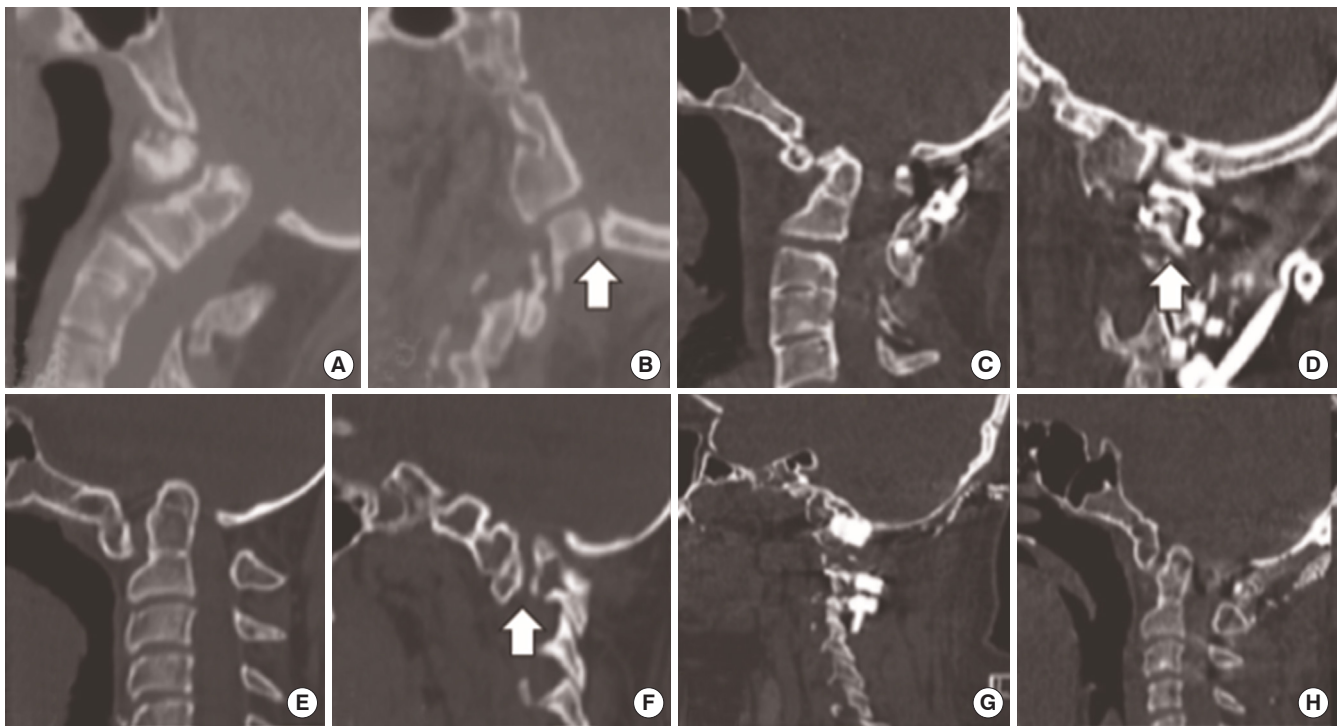


Fig. 5. This figure shows the 2 types of pseudo-joints described by author based on examination of computed tomography (CT) scans in over 200 patients. Type I (panels A-D) is characterized by partial articulation of proximal part of C2 pars with the inferior surface of the occiput (B). Hence, weight of the head is transmitted by both the true joint and pseudo-joint. As can be seen here (panels A-D), this patient presented with moderate to severe BI (A). CT scan (with coronal reconstruction) showed a type I pseudo-joint on one side (B, arrow) and normal joint on the other side (not shown in this figure). Here, a complete joint-remodeling was performed after which the spacer could be placed partially over the true and pseudo-joint (D, arrow). On the other side, the spacer was placed completely within the true joint (as there was no pseudo-joint). CT scan demonstrated a satisfactory reduction (C). The other type of pseudo-joint is type II, where the true joint is completely vertical, and the spacer has to be placed completely within the pseudo-joint (E-H). The patient in this example (E-H) as can be seen having a severe BI (E). There was presence of type II pseudo-joint on one side (F) and a type I joint on the other side (not shown here). As can be seen here, the true joint is completely vertical (F, arrow), and the whole weight of the head is completely supported by the pseudo-joint. The spacer was placed within the pseudo-joint (G) and a complete reduction was performed (extra-articular distraction with DCER type III surgery). DCER, distraction, compression, extension, and reduction.

tion (Fig. 6A, B). It is important to approach the side first, where the VA is in nondominant. If the VA is placed over joint (as seen in Fig. 6A), it should be dissected carefully and mobilized away from the joint (Fig. 6B). The entire dissection of this area should be performed under the microscope. The surgeon should always be ready to manage in case of inadvertent injury to the VA. In case of injury (Fig. 6C-E), the VA should be repaired using a 9-0 prolene under microscope.

7) Extent of fixation

Like any spinal deformity, the extent of fusion is dictated by the degree of complexity. Thus, for type I surgery, the author prefers a short segment fixation (occiput and C2 translamina, 2-point fixation). For type II & III surgeries, a long segment fix-

ation is preferred (3-point fixation), i.e., occiput-C2/C3 or occiput-C3/C4 fixation. However, as mentioned earlier, the author currently prefers to do a 3-point fixation for almost all cases.

8) Occipital fixation

All DCER procedures undergo an occipital fixation. The reasons include: (1) In all these cases, the C1 is occipitalized, hence the C1 and occiput form a continuous bony mass. (2) DCER is based on using the spacer as a pivot joint (type II lever, Fig. 7A). Hence, occipital screw placement allows a maximal length of lever arm, thus proportionately reducing the force at the screw bone interface. This enhances the strength of the construct fixation (Fig. 7B). In cases where occiput is fused with C1, occipital screw fixation is a safer and easier technique. Presence of VA

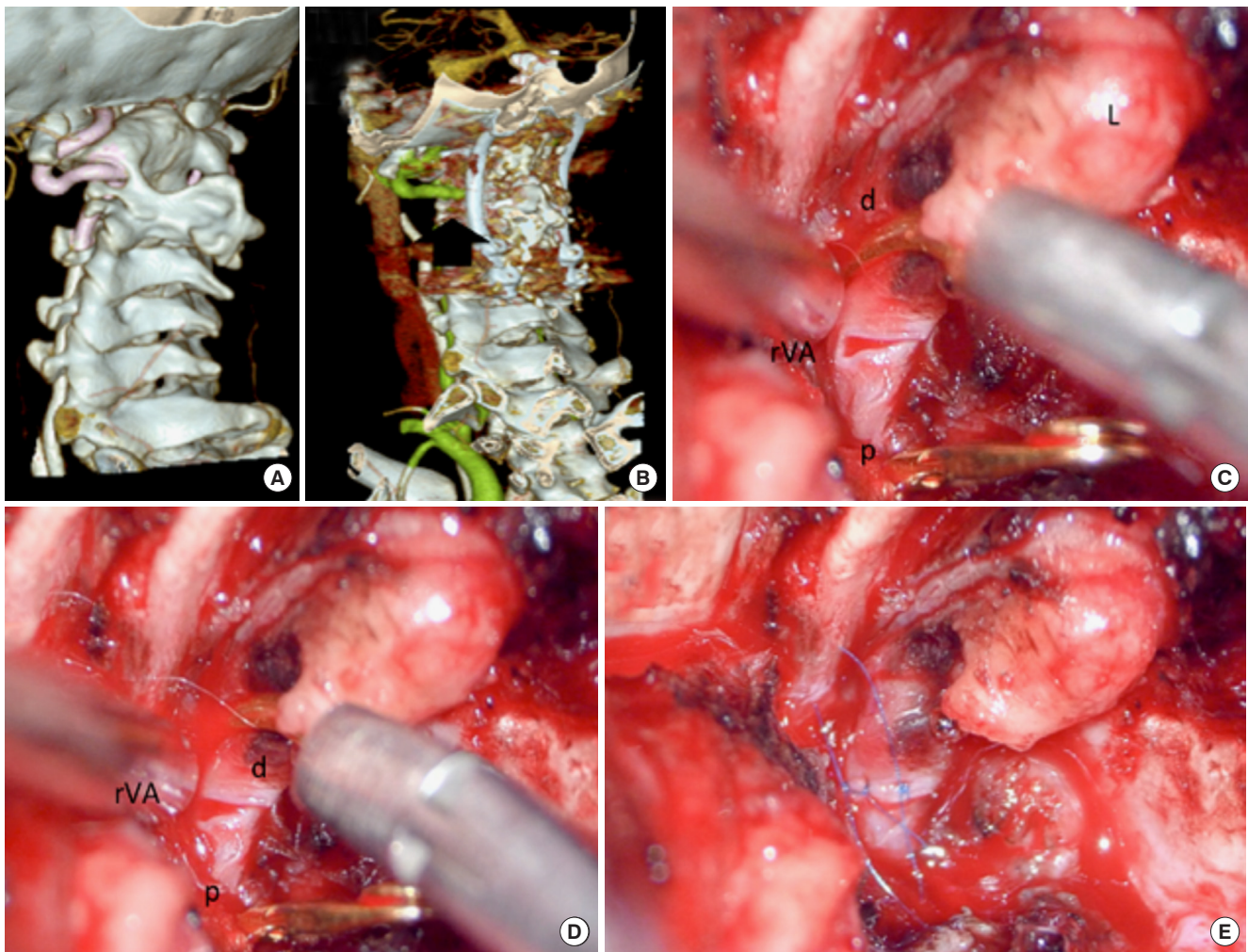


Fig. 6. Panel A shows location of an anomalous vertebral artery (VA) over the joint line as demonstrated excellently in 3-dimensional computed tomography angiogram. Panel B shows how VA has been mobilized for performing DCER (distraction, compression, extension, and reduction) and instrumented fixation. Panels C-E shows technique of repair of VA. Panel C shows the rent in the VA (right VA [rVA]) with temporary clips applied both proximally (p) and distally (d). Panel D shows that release of the distal clip (d) shows a good backflow through the rent indicating a good cross circulation. Panel E shows the repair of the VA with application of three 8-0 sutures. Following this, the VA was retracted superiorly and the exposure of the joint was continued. L, left side.

anomalies especially in cases with occipitalization of C1 arch, increases the risk of vertebral arterial injury for C1 screw placement in these cases.

9) Surgical Technique

As mentioned earlier, DCER stands for distraction, compression, extension and reduction. The technique is a caveat of the Archimedes lever principle (aka “picture of him lifting Earth using a long rod, with the fulcrum over a pebble- “Give me a place to stand and I will lift the Earth”). The ‘pebble’ is of course the spacer, the long lever is the distance of the rod from the spacer to the occiput and the ‘Earth’ is the head, which is pulled out

from the spine to correct BI and AAD.

10) Position and initial exposure

The patient is positioned prone. I do not use any overnight skeletal traction. I prefer to keep the patient on a horseshoe with plenty of eye padding to prevent any ocular injury due to pressure. Intraoperative skeletal traction may be used with mild weight (around 2 kg). Iliac crest bone graft is prepared.

11) Exposure of joints

Once the occiput, C2, C3, and C4 are exposed, the joint exposure is started. It is important to perform this technique un-

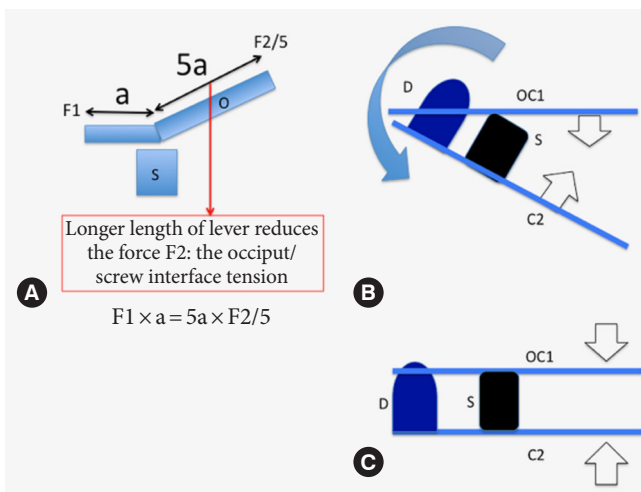


Fig. 7. Figure shows the fundamental principle of DCER (distraction, compression, extension, and reduction) and the lever mechanism justifying the placement of an occipital screw. Panel A shows the lever principle. ‘a’ represents the distance between the spacer (s) anterior border of occipito-C1–2 (OC1–2) osseous structures. F1 is the force required to stretch the ventral ligaments. ‘5a’ is the distance between the occipital screw and the spacer. It may be now seen that the entire construct forms a type II lever with the spacer in center (like a see-saw). Since the distance ‘5a’ is much longer than the distance ‘a’ (in this case, assumed to be about 5 times more), thus, the amount of force at the screw-bone interface will be about 5 times less (as noted in the formula given below). Panel B shows the fundamental mechanism of DCER. The spacer is placed first which leads to Distraction (“D”). The basilar invagination (BI) gets reduced to a great extent, but the atlanto-axial dislocation (AAD) does not get reduced much. Following compressive (“C”) forces between occiput and C2 (shown as arrows), this leads to compressive and extension (at the OC1–2 joint; “E”) which finally leads to reduction “R” of the AAD and also residual BI (C). Thus, the name DCER implies the entire surgical process which leads to complete reduction of AAD and BI.

der microscope. I prefer to use a variable impedance non-stick bipolar system (Vesalius or Sutter). This is particularly useful to prevent any bleeding and also to perform rapid dissection. Optimal saline irrigation is important. It is important to understand the points of venous bleeding. These include (1) epidural venous plexus medially, between the laminae and the dura: This is controlled by placing a thin pressed dry spongstan (gelfoam) between the dura and the lamina. (2) Venous plexus over the C2 ganglion lateral to the pars: There is presence of dense venous plexus lateral to the pars. It is thus important to prevent going into this region. If transgressed here, the bleeding may be controlled using spongstan packed with wet gauge for some

time. (3) The third area of venous bleeding is from venous plexus. Under the C1 arch posterior to the lateral mass. These are again epidural venous plexus, which drain from the epidural area into the vertebral venous plexus. These again may be controlled by keeping a wet spongstan gently pressed with a cotton for some time. *It is important to understand that surgical dissection in this area is generally bloodless if the principles of dissection and hemostasis are followed.* The C1–2 joints may be easily exposed by following the superior border of the C2 lamina laterally which directly leads to the joint. The C2 root ganglion may be next seen over the joint. I prefer to save the C2 root by retracting it superiorly. However, many times, especially in developmental cases, the root is ‘hypertrophic’ or is entrapped between the true and the pseudo-joint and will have to be cut. It is important to cauterize and then cut the root sharply. This should be done well away from dura to avoid any cerebrospinal fluid (CSF) leak from the dural sleeve.

12) Optimizing the joints

Once the joints are exposed, the joint surfaces should be optimized for placement of spacer (Fig. 8, Supplementary video clip 1). I prefer to use the term ‘optimizing’ as it encompasses all terms from just denuding the cartilage to joint remodeling. In cases where the SI is between 90°–100° (type I surgery), the joint surfaces are drilled gently using a diamond drill. It is again important to emphasize that this entire procedure should be performed under a microscope. In cases, where the SI is between 100°–160° (type II surgery), joint remodeling is performed. Here, the posterior margins of the joints are drilled to change the SI to around 90°. In cases where there is a presence of pseudo-joints (type III surgery), these are identified by placing a periosteal elevator under the occiput and the superior surface of the pars of C2 and turning it gently to separate the joints (Figs. 8-10, Supplementary video clip 1). In type III surgery, since the pseudo-joints are more dorsally located than the true joint (Fig. 5), the identification and its optimization is relatively easy. In many, I was surprised to see even a cartilage and synovial fluid within the pseudo-joint, suggesting a very early genesis of the pseudo-joint. It is also important to understand vertical joints (SI, 160°–180°) may be present even on one side. This may be responsible for rotary AAD and neck tilt to one side. Such cases require different height spacers to be placed on each side correct the neck tilt. Hence planning to place correct size of spacers is very important.

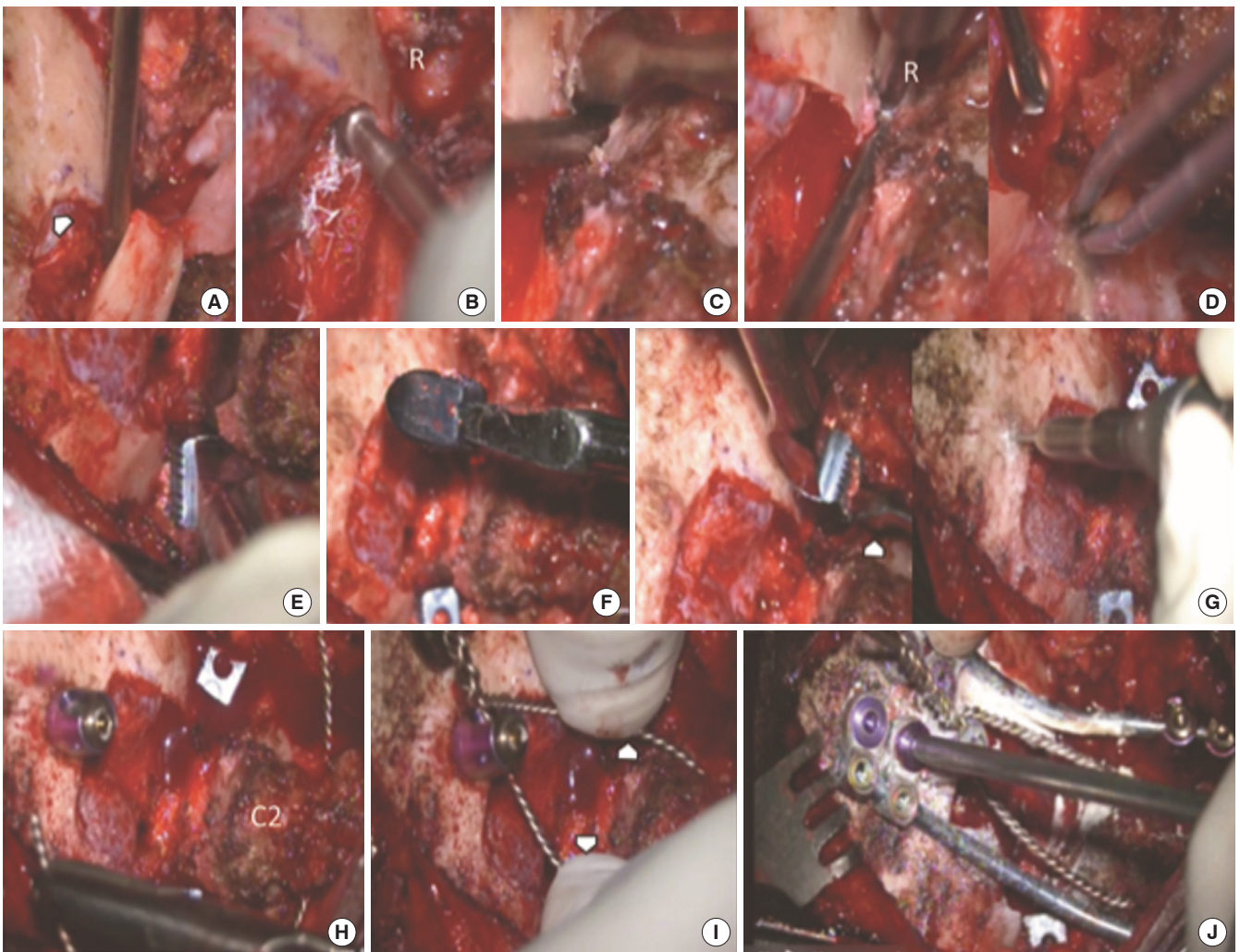


Fig. 8. Figure showing intraoperative pictures of the technique of DCER (distraction, compression, extension, and reduction). (A) Foramen magnum decompression. The author prefers to remove the margin of foramen magnum as a block so that the bone may be used for fusion. (B) The lateral margins of the foramen magnum are drilled to ensure that there is no compression along the sides of the foramen magnum. (C) The occipito-C1-2 (OC1-2) joint is then meticulously 'loosened' using a periosteal elevator. The periosteal elevator may be inserted into the joint and then turned to one side to free the joint. (D) The joint cartilage is denuded using a diamond drill. Following this the periosteal elevator may be inserted again to further free the joint surfaces. The same process is then repeated on the other side. Please note that in this case, both the sides are pseudo-joints, hence the joint is being accessed just under the occiput. (E) Once the joint surfaces are adequately loose on both sides, the correct sized spacer is then selected and inserted into the joint cavity. The figure shows insertion of the spacer on the left side. Panel F shows the profile of the spacer before being inserted into the right joint cavity. Panel G shows the right-side spacer being inserted. Please note that the convex curved surface of the periosteal elevator (arrow head) to 'rail road' the spacer into the joint cavity. (H) Next, a temporary screw is placed on the occiput along with the 'inne'. A braided stainless-steel wire is next passed under the C2 spine and through the occipital screw hole. (I) This occipital wire is then tightened over the occipital screw. The assistant has to press both the wires against the C2 lamina (arrowheads). The wire should be tightened slowly and gently to allow gradual compression of OC1 against C2 followed by extension of OC1-2 joint. This extension will then relieve the dural compression, which will be characterized by appearance of dural pulsations and some bleeding from the epidural space. (J) Once there is complete reduction, the same may be confirmed using an O-arm. Following this, OC3-4 fixation is being done. Alternatively, OC2-3 fixation can be done. But it is important to perform a 3-point fixation in all cases. Following fixation, the wire is cut and removed. The temporary screw is also removed and the hole is plugged with bone wax. R, right side.

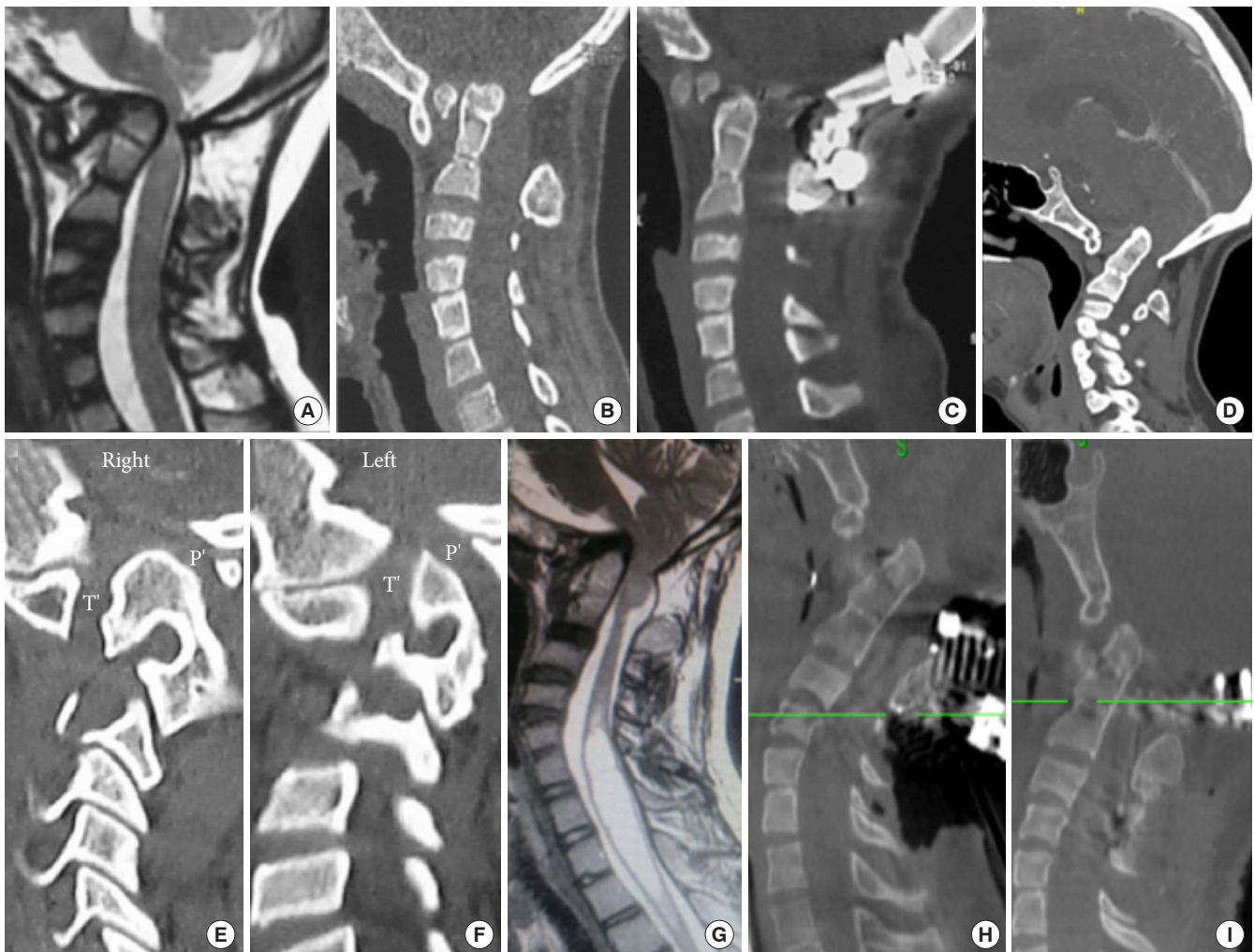


Fig. 9. Figure showing an example of a patient with Os-odontoideum with basilar invagination (BI) and atlantoaxial dislocation (AAD) (A, B) with severe cord compression in a 14-year-old boy. It may be seen here that the Os is placed ventral to the dens. (C) Following DCER (distraction, compression, extension, and reduction) a complete reduction and realignment has been achieved. The Os as can be seen here, has moved to a superior position. Panel D shows another example of a patient with severe BI and AAD. (E, F) The parasagittal images show that the true joint (T') is vertical, while the pseudo-joint (P') can be seen being formed between the superior and proximal surface of pars and the inferior surgery of occiput which is flattened out like a foot. (G) Magnetic resonance imaging shows severe cord compression with syringomyelia. (H) Following placement of 7-mm height of spacer on one side and 8-mm height on the other, the BI was corrected but there was still a significant space between the dens and the clivus. We then increased the height of the spacer to 8, 9 mm, respectively. (I) This leads to a good correction after performing compression and extension.

13) Placement of spacers

Spacer placement is one of the key steps of the surgery. It is like placement of an intervertebral implant or an arthroplasty device. Hence, it is very important to adequately prepare the joint surfaces before performing this step. The height of spacer is roughly 80%–90% the height of BI as calculated by Chamberlain line. The reason is that, following compression & extension, both AAD and also the residual BI gets corrected (Fig. 7). It is also important to understand that when a spacer is placed

on one side, the joint surfaces on the other side come closer to each other. This may make placement of spacer on opposite side difficult. For overcoming this, I prefer to use a 'rail-roading' technique by inserting a periosteal elevator and passing the spacer over it so that the elevator guides the placement of the spacer (Fig. 8G).

14) Drilling the posterior margin of occiput

This is an important step before performing compression

and extension (C&E component of DCER). I prefer raising a small bone flap from the margin of the foramen magnum (Fig. 8A). Earlier, I used to drill the margin of foramen magnum and used the bone dust for fusion. I prefer the drilling of rim of foramen magnum as the first step of the surgical procedure. It is also important to note that the posterior rim of occiput should be drilled to just 2–3 cm. Excessive drilling may precipitate tonsillar ptosis. In addition, it is important to drill laterally till the lateral margin of foramen magnum to prevent any lateral compression during compression and extension.

15) Compression and extension

This is the key step of DCER and consists of the compression

and extension component (Figs. 8-10, Supplementary video clip 1). I prefer to use a braided no. 20 stainless-steel (SS) wire tied between the temporary occipital screw and inferior border of the C2 spine. Gradual turning of the wire leads first to compression between OC1–2 tightly holding the spacer in place. Following optimal opposition between the spacer and the joint surfaces, it is next followed by extension at the OC1–2 joints. One may notice during this procedure, the dura starts becoming lax and starts pulsating indicating release of compression ventrally. It is very important to note that the procedure of compression and extension must be performed very slowly so as to allow gradual stretching of ventral ligaments. It is also important for the assistant to press both the wires against the C2

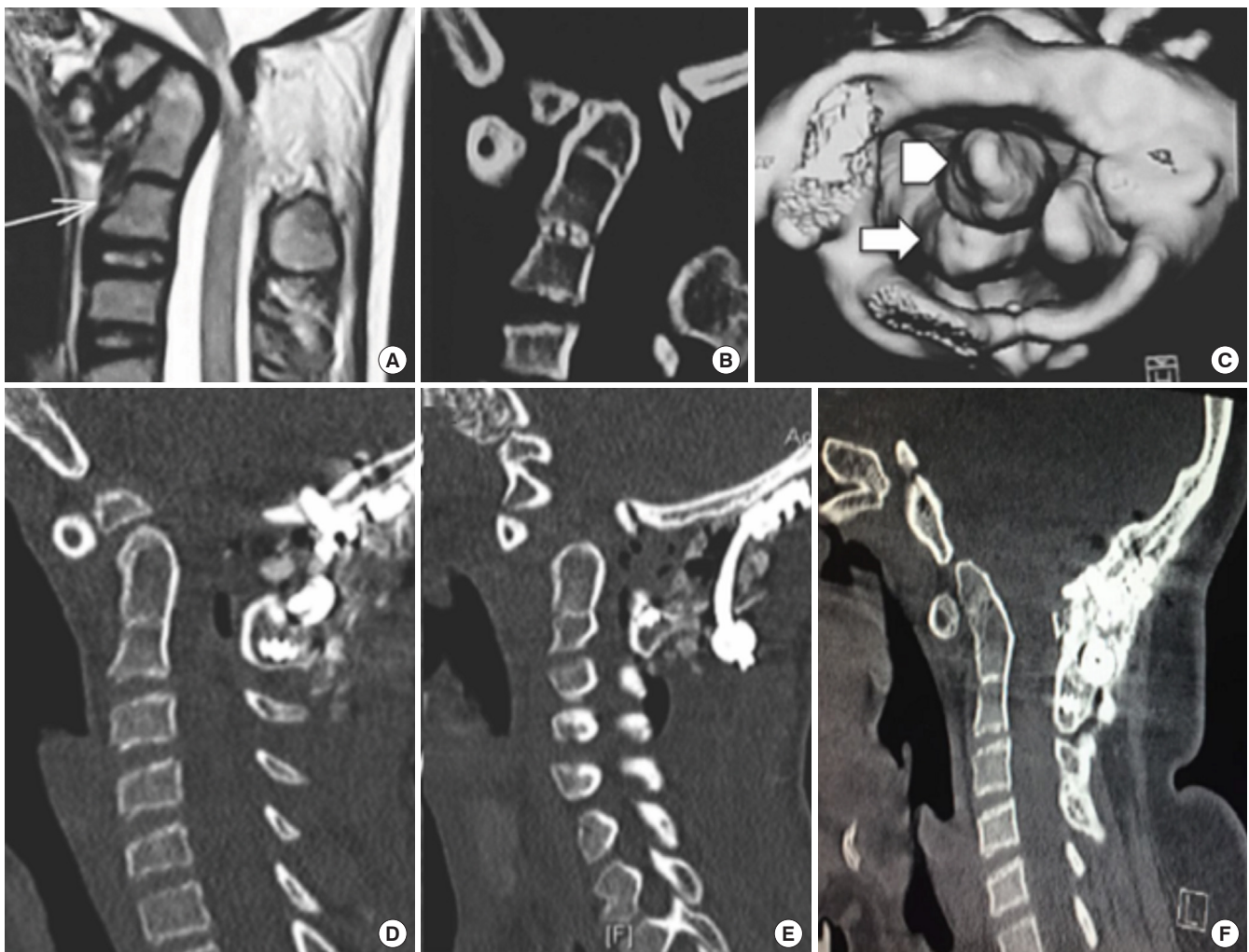


Fig. 10. Another example of DCER (distraction, compression, extension, and reduction) with a fractured Os-odontoideum (magnetic resonance imaging and computed tomography scans, A, B, and C, respectively). Arrows in panels A and C show the backward angulation of dens with the os being preserved in the normal position. (D) Following DCER, there is a complete correction. (E) Parasagittal image showing instrumentation. (F) Following 1 year after surgery, one may notice solid bone fusion posteriorly and also fusion of the os-odontoideum.

lamina (see Supplementary video clip). The extension between the OC1–2 joints is maximized when the occiput comes in contact (or can come as close as possible) with the C2 spine. I always prefer to take a CT using O-arm at this stage so that I can confirm complete reduction. If at this stage, if complete reduction is not seen, one may consider using a higher height spacers. I prefer to perform the entire procedure using motor evoked potentials (MEP) monitoring. However, in cases where the patients have significant neurological deficits, there may not be any waveforms recorded with MEP. In such situations, I prefer to perform a D2 wave recording by passing a thin epidural catheter.

16) Instrumentation

Following complete reduction, instrumented fixation is next performed (Fig. 8). I prefer to place 4 occipital screws fixed to a contoured rod with 4 clamps (VERTEX, Medtronic Sofamor Danek, Warsaw, IN, USA). In all cases, I prefer to perform a long segment 3-point fixation (OC2–3 or OC3–4). Following

this, the occipital and C2 spine margins are roughened using a diamond drill and bone graft is placed to enhance bony fusion.

7. Development of Customized Instrumentation

Over the years, we developed some unique instruments and implants. These are briefly described below:

1) Titanium C1–2 spacers

These are hollow bullet shaped titanium spacers of variable heights ranging from 4 mm to 22 mm. (Indian patent pending, number 201911002700 titled “Implant system to enhance the cranio-vertebral junction stabilization”). The system consists of an initial trial implant, which is a solid, smooth biconvex construct which is initially placed to create an optimal trajectory and space. This is followed by placement of the final implant which is a biconvex ‘bullet shaped’ hollow structure with serrated margins (Fig. 11). The hollow cavity is filled with autologous bone or bone mixed with hydroxyapatite. Both the trial implant and spacer are held by a special instrument holder which is

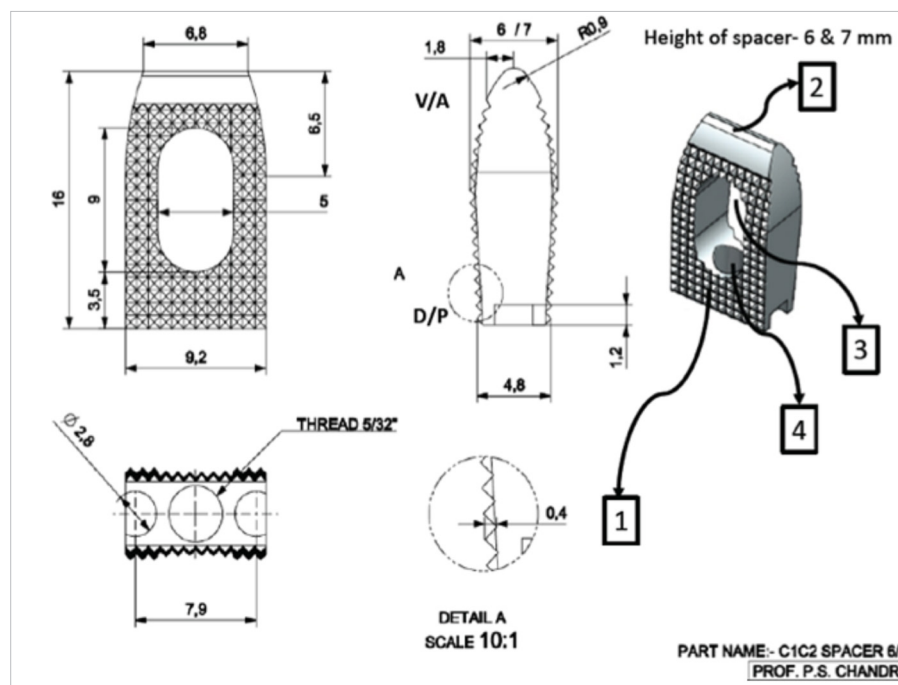


Fig. 11. The following shows the construct of the customized C1–2 spacer (patented) developed by author. It has the following advantages: The ventral end is tapered bullet shaped and this construct allows the construct to be introduced easily. The ventral end (2) is wedge shaped so that the spacer may introduced easily. The superior and inferior surfaces (1) are serrated so that the spacer can get a proper grip over the C1–2 joint surface. The central portion is a hollow core which may be filled with autologous bone or bone like material. The dorsal end (4) is used to attach a screw driver which may be used to hold and drive the spacer inside the joint cavity. The whole shape of the space is like a ‘bullet shape’ so that it may partly convert the joint into a pivot joint to allow the technique of DCER (distraction, compression, extension, and reduction) to be performed.

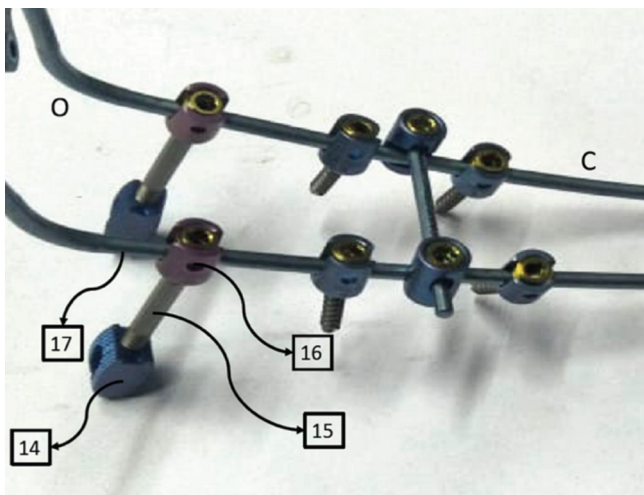


Fig. 12. This shows the unique concept of the 'spacer screw' (15). This screw attaches the spacer (14) to the occipito-cervical rod (15) via a screw head (16) which consists of an 'inne' and an 'outie.' This system allows the spacer to be rigidly retained within the C1–2 joint spacer and significantly reduces the chance of the spacer getting displaced (*patented*). O, occipital; C, cervical.

screwed to the hole along the posterior part of the spacer. The biconvex shape has 2 major advantages: (1) Firstly, it allows self-retraction of the joint surfaces and easy placement of the spacer. (2) The biconvex shape allows the joint to be converted into a 'pivot' joint which again allows the technique of DCER to be performed optimally. (3) There is special 'spacer screw' which may be screwed into the hole in the posterior part of the spacer and then the proximal part may be fixed to the rod of the occipito-cervical construct. The spacer screw is placed after the C3 and C4 lateral mass screws are placed, but before the rod placement so that the exact height of the spacer screw may be measured and placed (Fig. 12). Once, the spacer 'snuggly' fits into the joint, the holder is then removed. The placement of spacer constitutes the 'D' element of DCER which is the distraction. These implants come in various sizes (PSC spacers).

2) Universal craniovertebral junction reducer

The DCER technique using a temporary screw attached to the occiput addresses only 2-axis reduction, i.e., distraction and extension at OC1–2 joint. It cannot address the reduction along the sagittal plane (Fig. 13) i.e. it cannot 'push forward' the C2 to reduce the inherent C1–2 listhesis which is present in many cases. The C2 dorsal listhesis while minimal in some cases may be significant in many cases and cannot reduce unless an additional ventral 'push' is provided to C2. Furthermore, the lateral

mass screw and the wire may always pull out from occiput as they are temporary constructs and have not been optimized to be used for this technique.

To overcome all the above shortcomings, the author developed a Universal craniovertebral junction reducer (UCVJR; developed in collaboration with Medtronic). Following, the spacer placement, the UCVJR is applied to the occiput. The UCVJR (Fig. 13) consists of an occipital plate along with a pulley system which can tighten a specially made cable looped around C2 using a special DCER tensioner. This provides the movements of compression. In addition, a simultaneously C2 translator allows a forward movement of C2 by pushing a screw attached to a process over C2 laminae. Thus, simultaneous extension at the OC1–2 joint along with ventral (forward) translation of C2 leads to a complete reduction of the AAD and BI. The system has several advantages as compared to earlier described wire technique. (1) Firstly, the occipital plate is so constructed that it firmly fits over the occiput with 2 screws. (2) The SS cable and DCER tensioner has been specially designed with adequate strength so that it can allow excellent reduction without the cable snapping. It is also quite flexible unlike the braided SS wire which the author was using earlier. (3) The C2 translator is unique as it allows the forward/ventral motion of the C2 thus also reducing the C2 listhesis. (4) Both the extensions at OC1–2 joint and forward translation of C2 over C1 may be provided simultaneously. Thus, the UCVJR for the first time introduced the concept of 3-axis reduction i.e., Distraction (provided by spacers) followed by Extension at OC1–2 joint (provided by cable) and ventral translation of C2 (provided by C2 translator). The combination of all movements allows a complete reduction of AAD and BI.

Once reduction is performed, it is confirmed with imaging and following fixation, the whole UCVJR is removed. It thus cuts down the tremendous learning curve associated with the treatment of this complex pathology, and at the same time provides safe, efficient, and simple method for reduction (even for very severe BI and AAD).

Following reduction, with either the wire technique or with the use of UCVJR, instrumented fixation is then performed. The author generally prefers to use occiput–C2/C3 and/or C4 (3-point) fixation. This is in sync with the management of complex spine pathologies to perform a long segment fixation to allow better stability and provide better chances of long-term bone fusion.

Following instrumented fixation, the wire or the UCVJR is removed, complete hemostasis is obtained and closure without

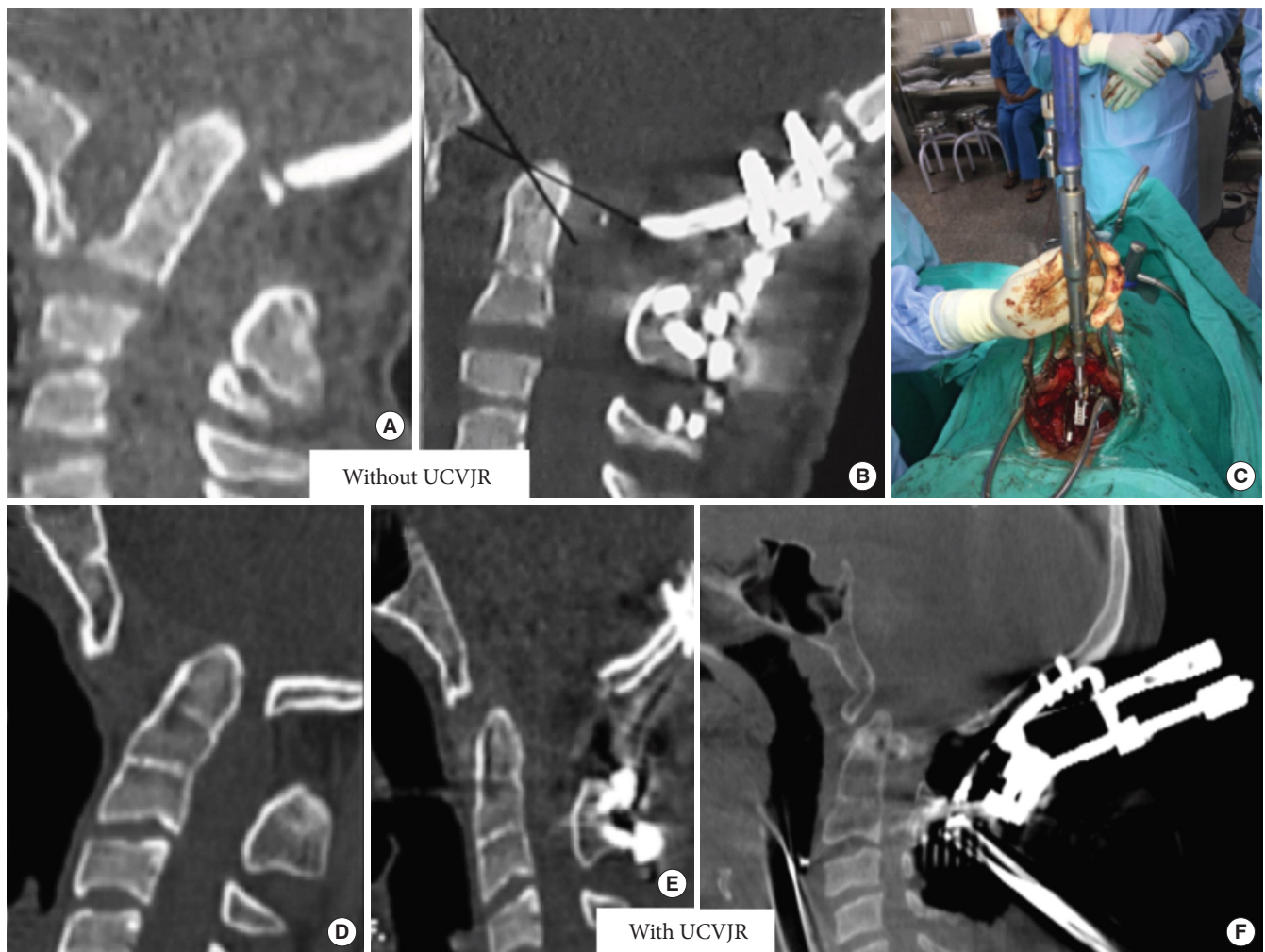


Fig. 13. This figure shows the utility of universal craniovertebral junction reducer (UCVJR). Panel A shows a very severe basilar invagination and atlantoaxial dislocation. The odontoid process as can be seen is tilted backwards and causing very severe spinal cord compression (B). Following DCER (distraction, compression, extension, and reduction) using a wire technique (see Supplementary video clip 1) the basilar invagination (BI) was corrected, but there was still a significant gap between the clivus and the dens. This is because, in severe BI there is also a posterior listhesis of the C2. Hence the center of gravity for both occipito-C1-2 (OC1-2) are not aligned. Hence, even after performing the DCER technique, the atlantoaxial dislocation may not get completely reduced. The UCVJR provides an additional axis of movement, i.e., a forward translation. Thus, for the case as shown in panel D, an optimal reduction was achieved using the UCVJR (C). Panel F shows the UCVJR in situ as seen intraoperatively using an O-arm. As it can be seen here, the distraction is provided by the spacers (as in DCER), followed a compressive force which is provided by a special tensioner along with a specially braided stainless-steel wire (with much higher break out force as compared to the atlas cable). As in DCER, continuation of the compressive forces will lead to the extension at OC1-2. Thus, for the first time, a mechanism has been created which can provide forces along all the 3-axis, i.e., distraction (D), followed by compression (C) which serves to lock the spacers properly), extension (E) at the OC1-2 joint and forward translation of C2 (T). Thus, use of UCVJR adds an addition “T” to the DCER making it DCETR.

drain is performed in layers. The author generally prefers to electively ventilate these patients overnight.

3) Postoperative management

I usually prefer to extubate in the type I surgery. For more complex type II and III surgeries, I prefer to electively ventilate them

overnight and get the patient extubated the next day. The patient should wear a Philadelphia hard collar for a period of 3 months. Isometric neck exercises are started within 1 week, as soon as the pain subsides to prevent disuse atrophy.

RESULTS

A total of 148 patients with average age 27.25 ± 17.43 years, ranging from 3 to 71 years (87 males) were operated.

1. Clinical Features

Patients presented with either or a combination of suboccipital pain (n = 48, 37.5%), restricted neck movements (n = 23, 17.9%), neck tilt (n = 40, 31%), quadriparesis (n = 114, 89%), sensory loss (n = 109, 85%), dysphagia (n = 5, 3%), bladder and bowel involvement (n = 16, 12.5%), drop attacks (n = 2, 1.5%), and respiratory difficulty (n = 2, 1.5%). Os-odontoidum was seen in 3 patients (2.3%).

2. Clinical Parameters

Using preoperative and post-operative Nurick grading 138 of 148 patients had improvement (93.2%). Mean preoperative Nurick grading was 3.13 ± 1.88 , which decreased to 1.24 ± 1.27 , $p < 0.0001$. Most of the patients reported subjective improvement on the day of surgery itself.

3. Pseudo-joints

Of 148 patients, 138 patients were able to trace preoperative CT in 276 joints by measuring both sides of the C1–2 joint (some patients had films which could not be uploaded into the hospital picture archiving and communicating system). Of these, 129 (47.4%) had absent (type 0) pseudo-joints, 52 (19%) had type I pseudo-joints, and 91 (33.6%) had type II pseudo-joints.

4. Coronal/Neck Tilt

This was present in 47 of 138 patients (32%). On CT examination, this was found significantly associated with the presence of pseudo-joint to one side, or difference of type of pseudo-joint (i.e., type I vs. type II) in between the 2 sides (Pearson correlation coefficient -0.658, $p < 0.0001$).

5. AAD

Mean preoperative AAD was 7.24 ± 2.74 , and postoperative was 3.56 ± 2.14 . ($p < 0.0001$).

6. Basilar Invagination

There were significant reduction of BI as assessed using Chamberlain line (6.44 ± 4.24 mm, $p < 0.0001$), McRae line (4.83 ± 4.11 mm, $p < 0.001$), Wackenheim clival canal lines (6.5 ± 3.79 mm, $p = 0.04$), and Modified Ranawat index (6.58 ± 4.18 mm, $p < 0.0001$).

BI severity correlated with increasing type of pseudo-joint using Chamberlain line (correlation = 0.317, $p = 0.001$), McRae line (correlation = 0.488, $p < 0.0001$), Wackenheim (correlation = 0.328, $p = 0.001$), and Modified Ranawat index (correlation = 0.276, $p = 0.006$).

7. SI, CI, and CCT with Pseudo-joint

There was a significant correlation of increasing SI with increased grade of pseudo-joint for both right (correlation = 0.734, $p < 0.0001$) and left side (correlation = 0.744, $p < 0.0001$). Increasing CI was also found to be significantly associated with increasing grade of pseudo-joints for both right (correlation = 0.644, $p < 0.0001$) and left sides (correlation = 0.553, $p < 0.0001$). This correlated with our study.^{1-4,17,18} Though CCT was not found correlating with increasing grade of pseudo-joints. However, there was a significant difference between the pre-operative and post-operative CCT values ($p < 0.001$).

8. Lordosis Correction

There was a significant decrease in the cervical hyper-lordosis after the surgery. Mean preoperative lordosis was $24.89^\circ \pm 18.51^\circ$, and postoperative was $15.38^\circ \pm 12.66^\circ$, which was statistically significant; $p < 0.0001$. There was a significant correlation of increasing grade of pseudo-joints with increasing degree of lordosis (correlation = 0.435, $p < 0.0001$). Patients with pseudo-joints have a higher degree of hyper-lordosis compared to patients with no pseudo-joints, i.e., $29.45^\circ \pm 18.74^\circ$ compared to $17.53^\circ \pm 15.85^\circ$ respectively ($p = 0.002$). Postoperatively, both groups had a significant decrease being $10.30^\circ \pm 8.73^\circ$ and $18.53^\circ \pm 13.75^\circ$ respectively.

9. VA Anomalous Course

Anomalous VA was found in 21 of 51 (41.4%) of patients. However, the VA was found actually over the joint surface in only 10 of 51 (20%). We found increased incidence of VA anomaly with pseudo-joint (46.85%) compared to normal joints (31.57%). However, this was not found statistically significant, $p = 0.38$.

10. Complications

There were 22 patients (14%) having complications. These included VA injury (n = 4; all had anomalous VA; 3 underwent primary repair; In 1 case, the rupture happened while inserting the spacer, hence the leak was large. Patient under postoperative angiogram and stenting), CSF leak (n = 6; required temporary lumbar drain in one patient; all cases resolved), pneumonia (n = 5; 2 patients underwent tracheostomy because of poor

respiratory effort), deterioration of power ($n=2$), implant infection ($n=2$; one patient needed to remove it), partial implant slippage ($n=2$; did not require repositioning), and extradural hematoma ($n=1$; no intervention required).

Mortality was seen in seven patients (4%). Causes of deaths were VA injuries ($n=2$), acute quadriplegia ($n=1$), pneumonia ($n=3$; all had severe spastic quadriparesis, bedridden, malnourished and poor respiratory effort), and myocardial infarction ($n=1$).

DISCUSSION

AAD along with BI usually occur in congenital conditions is a complex pathology and demands detailed knowledge and understanding of the CVJ anatomy.

The last decade has seen a paradigm shift to standalone posterior fixation from the traditional transoral odontoidectomy and posterior fixation.^{1-4,6,18-27} Goel's technique was no doubt the starting point of understanding the shift of this paradigm.^{6,9,20-23,26-34}

However, it is important to understand that Goel's technique is effective in conditions where C1 is not fused with occiput or in conditions where there is no BI with a posterior and upward tilt of the dens.^{3,4,17,18} In such cases, an additional axis of sagittal motion would be required to completely correct the deformity.

Normally, BI occurs in congenital cases with occipitalized C1 arch. In such cases, the dens usually prolapse into the C1 and is usually tilted upwards and backwards. Hence, just a distraction with a spacer may not be enough to correct the spinal cord compression and deformity.

Thus, to overcome this shortcoming, we introduced the technique of DCER.^{1-4,11,12,17,18} As described in the material and methods in detail, this technique for the first time provided a second axis of motion, i.e., extension at the OC1-2 joint, as a result of the continuation of a compressive force applied between the OC1-2 joint. This was achieved with a simple technique of applying a temporary screw over occiput and using a no. 20 stainless steel braided wire tightened between the occipital screw and the C2 lamina (Fig. 8, Supplementary video clip 1). This of course evolved over several years, where the author initially used rigid instrumentation to provide this mechanism² and then started using the SS wire.

We were also the first to demonstrate in the literature, the relationship between facet joint inclination and the severity of BI. To quantify this, we described new joint indices¹ which included SI and CI. Both, (especially the SI), correlated with the earlier described indices, and also the severity of BI and AAD. Based on this study, we established that increasing inclination of the

facet joints was responsible for increasing the severity of BI and AAD.

In a paper following that, we proposed a surgical strategy based on the degree of severity of SI.³ Thus, in mild joint inclinations ($SI < 100^\circ$), where it was easy to place the spacer, we suggested joint 'optimization' and spacer placement. In more severe cases (SI approximately between 100° - 160°), we suggested (again for the first time) the technique of joint re-modelling. This primarily meant drilling of the posterior lip of the inferior joint surface and anterior lip of the superior joint surface to make the joint surfaces 'parallel' and 'flat' and more important parallel to the ground surface with the patient being considered in standing position (i.e., SI is about 90°). In this paper, we also solved the problem of vertical joints which are known to occur in some very severe BI. While Goel et al.^{6,9,20-23,26-34} never mentioned them in his papers, it was by and large considered as a contraindication for a posterior only approach. For the first time, again we described the existence of pseudo-joints and also described the technique of how to identify them and place spacers within the pseudo-joint (extra-articular distraction with DCER). In the current paper, we have for the first time quantified the pseudo-joints into type I and type II depending on whether the true joint takes part in weight bearing mechanisms partially or completely. The incidence of pseudo-joints are surprisingly not rare in congenital anomalies (type I is 19% and type II is 33% in our series). In our study, the neck tilt correlated significantly with pseudo-joint, being more towards the higher type of pseudo-joint, i.e., if a patient has type I on one side and type II on another, then the neck tilt would be towards the side with type II. Similarly, if there was no pseudo-joint on one side and type I pseudo-joint on the other side, then the neck tilt would happen towards the side of pseudo-joint. This clearly indicates that pseudo-joints represent the side of greater instability and type II pseudo-joint have a greater instability than type I. Furthermore, this study has shown that a higher SI had a higher chance of formation of pseudo-joint. This again establishes the fact that increasing inclination leads to the formation of pseudo-joint as a compensatory mechanism to bear the weight of the head (that cannot be now transmitted through the true joint due to them being inclined or vertical). All these findings, we feel are of significant value for management and again have been described for the first time in literature.

The paper also demonstrated for the first time in literature, correction of lordosis following the technique of DCER. Mean pre-operative lordosis was $24.89^\circ \pm 18.51^\circ$, and postoperative was $15.38^\circ \pm 12.66^\circ$, which was statistically significant; $p < 0.0001$.

This clearly establishes the fact that DCER technique by correcting the deformity establishes correction of the whole spine curvatures.

VA anomalies are of concern, especially for surgeons who are not used to handling dissection of large vessels. We have found the incidence of the VA to be present in 41% of cases (in 21 of 51 cases), in patients who underwent CT angiogram with 3D reconstruction. Following some initial complications (n = 4), we started performing VA imaging in all patients. This not only helped us to identify the VA abnormalities, but also prepared us adequately for the case. It is important to perform this part of dissection always under a microscope with the vascular anastomosis set ready. It is important not to forget that if an anomalous VA exists, it would be always positioned ventral to the C2 ganglion. Hence, the C2 ganglion should be carefully dissected and separated from the VA, before cutting it. Sharp injuries may be usually handled by primary repair. Blunt injuries or injuries due to diathermy are difficult to handle. This is the reason, that meticulous dissection in this area is essential. It is also

advisable to dissect first on the side with non-dominant VA.

Our complication rate was 14%. CSF leak (n = 6) is usually transient and occurs mostly from the junction of the occipital screws and bone. To prevent this, we make a tap of 4-mm-thick over occipital bone and place 4.5-mm-thick screws. Such a strategy has allowed us to control this problem. If there is a dural tear, it should be sutured carefully under a microscope and glue should be applied. Most of our leaks subsided with re-enforcement sutures. In only one case, a CSF drain was required. Lower respiratory tract infection (n = 5) is a matter of concern, especially in patients who are bed ridden, malnourished and have a low respiratory reserve. One should not hesitate to perform a pre-emptive tracheostomy if the necessity so arises. We have seen deterioration of power in 2 cases. Both were early cases, and we were not able to obtain complete reduction. In 2 of our cases, there was a partial displacement of the implant. Again, both were in early cases, when we were not using customized implants. With the use of the customized bullet shaped (PSC) spacers, we have not had any case of implant displacement following sur-

Table 1. Summary of important findings of this paper and general points

1. Indication	The main indication of the DCER procedure includes congenital cases of BI and AAD with occipitalized C1 arch. The presence of a vertical occipito-C1-2 joint and anomalous VA present over the joint are not contraindications of this technique.
2. Imaging	Apart from MRI, CT scan with 3D reconstruction, sagittal midline and para sagittal joint reconstructions, and CT angiogram with 3D reconstruction are mandatory. Assessment of bone quality is also necessary (Vit D3 level, DEXA scan if required).
3. Measurement of Joint indices	These include sagittal inclination (SI: $87.15^\circ \pm 5.65^\circ$, approx. 90°), coronal inclination ($110.3^\circ \pm 4.23^\circ$) and craniocervical tilt (normal: $60.2^\circ \pm 9.2^\circ$; is increased in BI usually $> 80^\circ$)
4. Surgical planning	Primarily based on SI. If $SI < 100^\circ$, a DCER (with joint optimization only) only will usually suffice (type I surgery). For SI between 100° – 160° , a joint remodeling should be done prior to spacer placement (type II surgery). For $SI > 160^\circ$, it is important to identify the pseudo-joint and an extra-articular distraction and DCER is required (type III surgery)
5. Pseudo-joints	The paper for the first time describes 2 types of pseudo-joints. Type I pseudo-joint (19%): weight transmission partly supported by true joint and partly by pseudo-joint. Here, following joint optimization, the spacer could be placed over both true and pseudo-joint. Type II pseudo-joint (33%): Here the true joint is completely vertical, hence the spacer should be placed only within the pseudo-joint cavity. The higher SI in this study correlated with higher type of pseudo-joint.
6. Neck/coronal tilt	As shown in this study was always towards the more abnormal joint, i.e., type II $<$ type I $<$ no pseudo-joint.
7. Anomalous VA	Anomalous VA was found in about 41% of cases. However, the VA was present over the joint surface in only 20% of cases.
8. Customized instrument (UCVJR) and implants (PSC spacers)	Customized instrument (UCVJR) and implants (PSC spacers) were found to be safe, effective and easy to use. In addition, UCVJR was useful in providing an additional axis of motion (C2 forward translation)
9. Overall, the technique of DCER	Overall, the technique of DCER was found to be safe, effective, reproducible, quantifiable, in all spectrums of BI and AAD. The concomitant use of UCVJR and PSC spacers made it more effective, safe and are likely to cut down the learning curve.

DCER, distraction, compression, extension, and reduction; AAD, atlantoaxial dislocation; VA, vertebral artery; MRI, magnetic resonance imaging; CT, computed tomography; 3D, 3-dimensional; DEXA, dual-energy X-ray absorptiometry; SI, sagittal inclination; BI, basilar invagination; UCVJR, universal craniovertebral junction reducer.

gery. In addition, with the introduction of the spacer screw, we have attained very good fixation of the spacer to occipito-cervical contoured rod, virtually eliminating this complication.

Our mortality was 4%, and only in 2% it was related directly to surgery (VA injury on dominant side). Following the enhancement of our management and surgical techniques as described above, we have not seen any further VA injuries.

To allow the procedures to be performed more easily, we have developed a new instrument (UCVJR) and a customized C1–2 spacer implant.

The UCVJR prototype (developed in collaboration with Medtronic) has been successfully used in over 36 cases. As described above, it adds a 3rd axis of movement, i.e., forward translation of C2 thus aligning the center of gravity of C1 and C2 in the same sagittal axis. We feel that the device would be of great utility especially for young surgeons making the surgery safer, efficacious and easier, thus reducing the incidence of complications and cutting down the learning curve.

The customized 'bullet shaped spacers have been developed in collaboration with the dept of Biotechnology, Ministry of Science & Technology. These spacers (described in detail in material and methods) may be positioned easily, separates the joint surfaces (in view of the narrow anterior wedge shape), and allow the technique of DCER to be performed easily (in view of the biconvex shape) which helps to convert the joint into a type II lever (e.g., see-saw). Following the surgery, there is settling down of the joint surfaces, rigidly impacting the joint to the entire surface of the spacer.

The study lacks a control arm to compare the results. However, the study established safety and efficacy similar to the previous ones published by us and demonstrated certain important new findings.

CONCLUSION

The above study represents the evolution of techniques and methods in the development of treatment of CVJ anomalies. For me (PSC), it has been a revolutionary journey starting from a period where preoperative traction+transoral excision of odontoid+posterior fixations was being performed to current scenario where most complex anomalies may be treated through a single posterior approach only and in many cases, the surgery may be completed in a relatively short time. DCER in our experience has been found to be safe, effective, reproducible, and quantifiable. The above study has described our consolidated experience. Summary of main findings are described in Table 1.

CONFLICT OF INTEREST

The authors have nothing to disclose.

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SUPPLEMENTARY MATERIAL

Supplementary video clip 1 can be found via <https://doi.org/10.14245/ns.1938174.087>.

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