Techniques in the treatment of craniovertebral instability

Atul Goel, Praveen Sharma, Nitin Dange, Arvind G. Kulkarni

Department of Neurosurgery, K.E.M. Hospital and Seth G.S. Medical College, Parel, Mumbai - 400012, India

The techniques of craniovertebral region stabilization introduced and used by the senior author over the last 20 years are summarized. The lateral masses of atlas and axis are strong and largely cancellous in nature and can be used for direct implantation of screws. Opening up of the joint and placement of bone graft within the joint stabilizes the region and provides a large area for bone fusion. Distraction of the facets provides an opportunity to treat a range of congenital craniovertebral anomalies. The technique of exposure of the lateral mass of the atlas and axis and the atlantoaxial joint is technically relatively complex and needs precise understanding of anatomy of the vertebral artery and training with cadavers.

**Key words:** Atlantoaxial dislocation, basilar invagination, vertebral artery, syringomyelia

The treatment of patients with atlantoaxial instability is a surgical challenge and achieving a successful outcome for these patients is gratifying. The complications of surgery, however, are potentially lethal. Various methods of fixation have been described and used successfully in the treatment of patients with atlantoaxial instability. The techniques of craniovertebral fixation evolved during the 20th century as the anatomy and biomechanics of the craniovertebral region became clearer. In 1994, we suggested an alternative plate and screw technique of fixation of the lateral masses of the atlas and axis vertebrae[1] and later discussed our 14-year experience with 160 cases with mobile and reducible atlantoaxial dislocation managed by this technique.[2] The recent popularity of our technique and analysis of biomechanical and anatomical issues by a number of authors has prompted this publication.

**Surgical anatomy**

The course of the vertebral artery, extensive venous channels in the region, relationships of C2 ganglion, atlantoaxial joint and the dural tube and the normal and abnormal alignments of the region has to be understood prior to surgery on the basis of all the available radiological information. Our belief is that experience with cadaveric dissections is mandatory for successful conduct of the surgery.

The C1 and C2 vertebrae are called ‘atypical vertebrae’ and have unusual shape and architecture and a complex and important vertebral artery relationship. Injury to the artery during surgery can lead to catastrophic intra-operative bleeding and compromise to the blood flow can lead to unpredictable neurological deficits, which will depend on the adequacy of blood flow from the contralateral vertebral artery.

The vertebral artery adopts a serpentine course in relationship to the craniovertebral region. The artery has multiple loops and an intimate relationship with the atlas and axis bones. We observed a wide variability of the course of the artery in our specimens. The shape, size and location of the vertebral artery groove on the inferior aspect of the superior articular facet of the C2 and over the posterior arch of the atlas had wide variations. The vertebral artery during its entire course is covered with a large plexus of veins. The venous plexuses are the largest in the region lateral to the C1-2 joint.

After a relatively linear ascent of the vertebral artery in the foramen transversarium of C6-to C3, the artery makes a loop medially towards an anteriorly placed superior articular facet of the C2 vertebra, making a deep groove on its inferior surface. The extent of medial extension of the loop varies. The distance of the artery from the midline of the vertebral body of C2 as would be observed during a transoral surgical procedure is on an average 12 mm.[3] The vertebral artery loops away from the midline underneath the superior articular facet of the C2.

The dens or the odontoid process is flanked by two large, superior facets, extending laterally on to the adjoining pars-interarticularis and articulating with the inferior atlantal facets. Superior facet of C2 vertebra differs from the facets of all other vertebrae in two important characters, which make this region prone to vertebral artery injury during screw fixation. First is that the superior facet of C2 is present in proximity to the body when compared to other
facets which are located in proximity to the lumina. The second is that the vertebral artery foramen is present partially or completely in the inferior aspect of the superior facet of C2, while in other cervical vertebrae, vertebral artery foramen is located entirely in relationship with the transverse process. Unlike superior facets of all other vertebrae, they do not form a pillar with the inferior facets, being considerably anterior to these. The pedicle of the C2 vertebra is relatively small. The course of the vertebral artery in relationship to the superior articular facet of the C2 makes its susceptible to injury during transarticular and inter-articular screw implantation techniques. It was observed that the screw implantation in the superior facet of the C2 vertebra has to be sharply medial and directed towards the anterior tubercle of the C1 for trans-articular fixation and towards the vertebral body of C2 for interarticular fixation. As discussed in our previous paper on this subject, the pars-interarticularis can be divided into nine quadrants. The superior and medial compartment can be used for inter-articular technique of screw implantation. The average distance of the artery from the ganglion was 7.5 mm. It suggests that the dissection around the lateral end of the ganglion should be carefully done and under vision.

The inferior facet of the atlas is almost circular in most of the vertebrae without any significant difference in the mean anteroposterior and transverse (15 mm) dimensions. The thickness of the inferior facet under the posterior arch of the atlas is on an average 3.5 mm. The thickness of the posterior arch of the atlas separating the vertebral artery groove from the inferior facet is about 3.5 mm.

Operative technique for lateral mass plate and screw fixation [Figure 1]: Cervical traction is set up prior to induction of anesthesia and the weights are progressively increased to approximately 5-8 kilograms or one-sixth of the total body weight. The patient is placed prone with the head end of the table elevated to about 35 degrees [Figure 2]. Cervical traction stabilizes the head in an optimally reduced extension position and prevents any rotation. The traction also ensures that the weight of the head is directed superiorly towards the direction of the traction and the pressure over the face or eyeball by the headrest is avoided. Elevation of the head end of the table, which acts as a counter traction, helps in reducing venous engorgement in the operative field. The suboccipital region and the upper cervical spine are exposed through an approximately 8-cm longitudinal midline skin incision centered on the spinous process of the axis. The spinous process of the axis is identified, and the attachment of paraspinal muscles to it is sharply sectioned. The large second cervical ganglion is about 3.5 mm. It is first exposed widely and then sectioned sharply. This procedure provides a wide exposure of the lateral masses of the atlas and axis. Bleeding from the large venous sinuses in the region and in the extradural space can be troublesome. Packing of the region with Surgicel and gelfoam can assist in the control of venous bleeding. The joint capsule is cut sharply, and the articular surfaces of the joint are exposed. The adjacent synovial articular surfaces of the atlantoaxial joint are decorticated widely with a microdrill, and pieces of bone harvested from the iliac crest are stuffed into the joint space. The lateral aspect of the lamina and a part of the pars of the axis are drilled to make the posterior surface of the lateral mass of the axis relatively flat so that the metal plate can be placed snugly and parallel to the bone. Drilling also helps in reducing the length of the plate and in placing the screw more superiorly and almost directly into the lateral mass of the axis. Actual vertebral artery exposure is unnecessary either lateral to the pars of the axis or superior to the arch of the atlas.

Screws are implanted into the previously created guide holes in the lateral mass of the atlas and axis through a two holed (approximately 2 cm in length) metal (stainless steel or titanium) plate [Figure 4]. First, a screw is placed into the atlas. It is directed at an angle of approximately 15° medial to the sagittal plane and 15° superior to the axial plane. The preferred site of screw insertion is at the centre of the posterior surface of the lateral mass, 1 to 2 mm above the articular surface. Whenever necessary, careful drilling of the inferior surface of the lateral aspect of the posterior arch of the atlas in relation to its lateral mass can provide additional space for the placement of the plate and screw implantation. The screw may even be implanted by choosing an insertion point on the articular surface of the lateral mass of the atlas. Such a site is useful more frequently in children than in adult patients.

Screw implantation in the axis is relatively unsafe, because the intimacy of vertebral artery relationships. The preferred site of screw implantation in the lateral mass of the axis is in the medial and superior third. The direction of screw implantation must be sharply medial and superior and should be toward the superior aspect of the body of the axis vertebra towards the midline. The medial surface of the pars of the axis is identified before the implantation of the screw. The screw is directed at an angle approximately 25 degrees medial to the sagittal plane and 15 degrees superior to the axial plane. The angle of screw insertion varies, depending on the local anatomy and the size of the bones. The quality of cancellous bone in the lateral masses of the atlas and axis in the proposed trajectory of screw implantation is generally good, providing an excellent purchase of the screw, and avoids the vertebral artery.

The screws used are 2.9 mm in diameter in the adult patients and 2.7 mm in diameter in the pediatric patients. The length of the required screw is calculated on the basis of the size of the lateral masses observed on the preoperative radiological studies. The approximate lengths of the atlas screws are 20 mm in adults and 17 mm in children. The screws in the atlas and axis were almost similar in their length. The lateral masses of the atlas and axis are firm and cortical in nature, and, although preferable, it is not mandatory that the screws engage both the posterior and anterior cortices. If the screw traverses beyond the anterior cortex, it will lie harmlessly in the anteriorly displaced soft tissue because no critical neurovascular structures are in close proximity to it. In such a situation, however, there is a possibility of injury to the pharyngeal wall. Intraoperative fluoroscopic control was found to be helpful but not essential in determining the state of the screws. Large pieces of cortico-cancellous bone graft from the iliac
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Figure 1: Line drawing showing the lateral mass plate and screw fixation technique

Figure 2: Line drawing showing the patient position. The patient is placed under cervical traction and the head end of the table is elevated

Figure 3A: Line drawing showing the relation of second cervical ganglion to the vertebral artery and atlantoaxial facet joint

Figure 3B: Cadaveric dissection of an injected specimen showing the relationship of the vertebral artery and the C2 ganglion to the atlantoaxial joint

Figure 4A: Lateral film of X-ray of the craniovertebral junction with the head in flexion position showing atlantoaxial dislocation

Figure 4B: Lateral X-ray with the head in extension position showing complete reduction of the dislocation
Figure 4C: Post-operative X-ray showing reduction and fixation using C1 and C2 lateral mass plate and screw fixation.

Figure 5: Line drawing showing ‘double insurance fixation’ technique. The C1 screw is placed directly in the lateral mass of the atlas and the C2 screw traverses in a transarticular fashion from the lateral mass of the C2 to the lateral mass of C1.

Figure 6: Line drawing showing the procedure of atlantoaxial lateral mass distraction and fixation. Spacers are placed within the atlantoaxial joint.

Figure 4D: Axial image showing the trajectory of C2 pedicle screws.

Figure 7A: CT scan showing basilar invagination, os-odontoideum, occipitalisation of the atlas and fixed atlantoaxial dislocation.

Figure 7B: Post-operative scan showing reduction of the basilar invagination and realignment of the craniovertebral junction.
bone are then placed over the adequately prepared posterior elements of atlas and axis. After the wound is closed, cervical traction is discontinued. The patients are mobilized as soon as possible and advised to wear hard cervical collar for 3 months.

Complication avoidance: The most dreaded complication of the procedure is injury to the vertebral artery. Appropriate anatomical information of the region in general and in the case in question is the only way one can possibly avoid this eventuality. The vertebral artery can be injured during the process of lateral dissection of the C2 ganglion. The other potential site of injury is during the insertion of the screw in the axis. In the later situation, to control the bleeding, one has to pack the bleeding bone hole with bone wax. One can then prepare for an alternative site of screw insertion or use an alternative method of atlantoaxial fixation. Respect and care of all neural and vascular tissues and employment of precise technique are critical to success.

This technique helps in manipulating atlas and axis independently by obtaining fixation points in their strongest elements and hence has very versatile applications.

Double insurance fixation: We have recently introduced an alternative method of atlantoaxial fixation that combines transarticular method of fixation and the interarticular fixation technique. The technique combines the biomechanical strengths of both the more commonly used techniques of fixation and provides maximal stability to the implants [Figure 5].

Atlantoaxial joint distraction is emerging as a rational form of treatment for a variety of craniovertebral anomalies that include select cases with basilar invagination, fixed atlantoaxial dislocation, torticollis and syringomyelia. The most important component of the success of this treatment is appropriate case selection. The technique of conduct of the operation is significantly complex and appropriate training and experience is mandatory.

Technique of atlantoaxial joint distraction: The facets of the joint on both sides are distracted using a combination of varying sizes of osteotomes and customised spreader similar to the one used in the anterior cervical discectomy. Bone graft harvested from the iliac crest is packed in titanium metal spacers and used as a strut in the prepared atlantoaxial facet joints [Figure 5]. Reduction of the dislocation and of basilar invagination is evaluated by intraoperative radiographic control. The size of the spacers used depends on the space available within the distracted joint space as well as the amount of distraction required to reduce the basilar invagination. The average sized spacers measured 10 mm in length, 8 mm in breadth and 4 mm in height. Metal spacer had a single large or multiple small holes to assist in bone fusion and is tapered at one end to assist placement in the joint. Bone graft was stuffed in the distracted joint space in multiple pieces on all the sides of the spacer.

Indications for atlantoaxial joint distraction

1. Fixed atlantoaxial dislocation: Atlantoaxial dislocation has been described as ‘fixed’ or ‘irreducible’ when there is no

![Figure 8A: CT scan showing basilar invagination, fixed atlantoaxial dislocation and assimilation of the atlas](image1)

![Figure 8B: Postoperative scan showing reduction of the basilar invagination](image2)

![Figure 8C: CT scan showing the unusual slant of the atlantoaxial joint](image3)

![Figure 8D: Postoperative scan showing realignment of the facets of atlas and axis](image4)
radiographic reduction of the dislocation on full neck extension or after institution of cervical traction. Fixed atlantoaxial dislocation can be congenital in nature or can be secondary to trauma to the region. Congenital os odontoideum and fracture at the base of the odontoid process are frequent accompaniments of fixed atlantoaxial dislocation. Various authors have suggested a transoral decompression followed by a posterior fixation as the safest method of treatment of this complex anomaly. Treatment by posterior decompressive procedures has been reported to be associated with high complication rate. Some authors have reported success with a transoral decompression of the region, without any posterior fixation.

We observed that direct facet joint distraction results in reduction of the fixed dislocation in a significant number of cases [Figures 6-8]. With our experience, we feel that there may be a place for reduction of the ‘fixed’ atlantoaxial dislocation and a subsequent fixation, without the removal of any bony spinal element.\(^5\)

2. Basilar invagination: Basilar invagination was divided into two groups. In Group A basilar invagination there was a ‘fixed’ atlantoaxial dislocation and the tip of the odontoid process ‘invaginated’ into the foramen magnum and was above the Chamberlain line,\(^5\) McRae line of foramen magnum\(^7\) and Wackenheim’s clival line.\(^8\) The distance between the tip of the odontoid process and the posterior limit of the anterior arch of the atlas or the lower end of the clivus was at-least 3 mm in these cases. Patients with Group A basilar invagination constituted approximately 60% of all cases of basilar invagination in our series.

Majority of patients with Group A basilar invagination had a history of minor to major head injury prior to the onset of the symptoms. The pyramidal symptoms and affection of the kinesthetic sensations formed a dominant component. Spinothalamic dysfunction was less frequent. Neck pain as a major presenting symptom was in approximately 75% cases. Torticollis was present in approximately 50% cases. The analysis of clinical features suggested that the symptoms and signs in this group of patients were a result of brainstem compression by the odontoid process. The radiological features suggested that the odontoid process resulted in direct compression of the brainstem. Analysis on the basis of Chamberlain’s line showed that the basilar invagination was mild to severe in these cases. Modified omega angle measurements suggested that the odontoid process had tilted towards the horizontal rather than rostrally.\(^8\) The patients had a ‘fixed’ atlantoaxial dislocation and no atlantoaxial mobility could be identified on dynamic radiology in any case.

The standard and most accepted form of treatment of Group A basilar invagination is a transoral decompression. Majority of authors recommend a posterior occipitocervical fixation following the anterior decompression. It appears to us that the atlantoaxial joint in such cases is in abnormal position as a result of congenital abnormality of the bones and progressive worsening of the dislocation is probably secondary to increasing ‘slippage’ of the atlas over the axis. The slip of atlas over the axis appears to be accentuated by the event of trauma.

We had earlier attempted to reduce basilar invagination by performing occipitocervical fixation following institution of cervical traction.\(^5\) However, all the four cases treated in this manner subsequently needed transoral decompression as the reduction of the basilar invagination and of atlantoaxial dislocation could not be sustained by the implant. Wide removal of atlantoaxial joint capsule and articular cartilage by drilling and subsequent distraction of the joint by manual manipulation provided a unique opportunity to obtain reduction of the basilar invagination and of atlantoaxial dislocation. The joints were maintained in a distracted and reduced position with the help of bone graft and spacers. The subsequent fixation of the joint with the help of interarticular screws and a metal plate provided a biomechanically firm fixation.\(^9\) [Figures 7,8]

With our experience in handling the atlantoaxial joints in this group of patients, we have realized that the joint is not ‘fixed’ or ‘fused’ but is mobile and in some cases is hypermobile, and is probably the prime cause for the basilar invagination. The history of trauma preceding the clinical events, predominant complaint of pain in the neck and the improvement in neurological symptoms following institution of cervical traction suggests ‘vertical’ instability of the craniovertebral region.

The fixation was seen to be strong enough to sustain the vertical, transverse and rotatory strains of the most mobile region of the spine. Following surgery, the alignment of the odontoid process and the clivus and the entire craniovertebral junction improved towards normalcy. The tip of the odontoid process receded in relationship to the Wackenheim’s clival line, Chamberlain’s line and MCrae line suggesting reduction in the basilar invagination. The posterior tilt of the odontoid process, as evaluated by modified omega angle, was reduced after the surgery. We could obtain varying degrees of reduction of the basilar invagination and atlantoaxial dislocation. The extent of distraction of the joint and the subsequent reduction in the basilar invagination was more significant in younger than in older patients.

3. Selected cases of syringomyelia: The complex of basilar invagination, Chiari 1 malformation and syringomyelia is relatively common and there are multiple reports on the subject. Association of fixed atlantoaxial dislocation in the later group is less common but not rare. Such cases are generally treated by either anterior transoral or posterior foramen magnum bony decompression. The indications and need for opening of the dura and manipulation of arachnoid membrane, tonsils and obex and draining of the syrinx cavity are currently under debate.

It was observed that cases of syringomyelia where there was ‘fixed’ atlantoaxial dislocation with or without the association of basilar invagination and Chiari malformation constituted a discrete pathologic group. It was observed that in this group, the patients were relatively young, neck pain formed a part of the symptom complex and the motor symptoms and ataxia was far more prominent symptom. We observed that an attempt could be made to realign the bones in the craniovertebral junction in these cases, without resorting to any bony or dural decompression or neural
manipulation of any kind [Figure 9]. The fact that there was a remarkable clinical improvement following the reduction of the atlantoaxial dislocation and of basilar invagination makes it appear that the complex of atlantoaxial dislocation, basilar invagination and syringomyelia are probably secondary to the primary craniovertebral instability.

4. Treatment of torticollis: Torticollis is a common feature in all forms of basilar invagination. No specific treatment modality has been described to treat this symptom. Transoral decompression or a posterior fixation procedure can provide decompression of the region and fixation, but the torticollis remains untreated. We observed that reduction of the atlantoaxial dislocation and of basilar invagination by manual distraction of the atlantoaxial joint present an ideal opportunity for reduction of torticollis [Figure 10]. Although, differential distraction of the joints was not attempted in our patients, but this procedure may provide an additional reduction of torticollis.

5. Treatment of basilar invagination and atlantoaxial dislocation in cases with rheumatoid arthritis: Basilar invagination is commonly associated with atlantoaxial dislocation and the complex results in a significant degree of neck pain and myelopathy adding considerably to the disability secondary to affection of other joints. We recently reported the feasibility of craniovertebral region bone alignment, distraction of the facets of atlas and axis and direct lateral mass plate and screw atlantoaxial fixation for management of both basilar invagination and atlantoaxial dislocation secondary to rheumatoid arthritis. Our operation of craniovertebral realignment and stabilization without any bone decompression could be successfully employed in cases with atlantoaxial dislocation in the presence or absence of retro-
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odontoid pannus and in cases with basilar invagination. The patients showed a remarkable and sustained neurological and radiological improvement. However, it was not possible to assess the extent of regression of pannus in our cases as stainless steel metal plates were used for fixation.

In selected cases, only atlantoaxial joint distraction using bone grafts and spacers can be used for treatment of both basilar invagination and atlantoaxial instability. The spacers stabilize the region, distract the lateral masses and jam all the movements occurring at the joint. The methods employing the use of plates, screws or wires can all be avoided by this technique.

**Occipitalization of the atlas and craniocervical instability**

'Occipitalized' atlas or 'assimilation' of atlas is relatively common. Occipitalization or assimilation of the atlas is a commonly seen feature in congenital craniovertebral anomalies and represents a failure of segmentation of the basal occipital sclerotome and first spinal sclerotome. Occipitalization of the atlas is frequently associated with maldevelopment of the occipital bone, reduced length of the clivus and platybasia, occipital condylar and adjoining bone hypoplasia and non-formation or inadequate formation of the occipitoaxial joint. Fusion of the atlantoaxial joint and C2-3 spinal elements and a range of Klippel Feil spinal abnormalities are frequently associated. Occipitalization of the atlas is usually associated with basilar invagination and compression of the cervicomedullary cord by the odontoid process. Occipitalized atlas is usually associated with fusion anomalies of adjoining bones in the region and with basilar invagination.\[12–15\]

Although all authors do not uniformly agree, the craniovertebral region in such cases has been reported by some to be 'potentially' unstable.\[15-18\] Mobile and reducible atlantoaxial dislocation in the presence of occipitalized atlas is not common.

**Occipitocervical fixation:** A variety of methods of occipitocervical fixation in the presence of assimilation or occipitalization of the atlas have been discussed. These include the use of the occipital squama for wire or screw fixation of the occipital end of the implant (plate, rods or metal loops); and the posterior elements of the axis or lower cervical vertebrae for fixation of the caudal end of the implant. Jain et al\[19\] have described the technique of drilling occipital bone close to the foramen magnum and formation of an artificial arch of the atlas, which is subsequently used for atlantoaxial fixation. Although occipitocervical fusions have been performed for more than 50 years using various types of instrumentation and each type having their own advocates, it is fair to say that an ideal universally applicable technique has not evolved.

The stabilization of the axial end of the implant has been carried out using sublaminar wires and wires around and through the spinous process. In the year 1994,\[1\] we described the use of lateral mass of the axis and of atlas for screw implantation for stabilization of the cervical end of the occipitocervical plate (Figure 11). The occipital end of the plate could be fixed with the help of occipital screws or wires. In the year 2004, we discussed the feasibility of direct implantation of screws into the spinous process of the axis.
for fixation of the cervical end of the plate.\textsuperscript{[20]} The relatively strong and sturdy spinous process of the axis in cases with occipitization of the atlas facilitated screw implantation.

In the presence of fusion of the atlas with the occipital bone, occipitocervical fixation for stabilization of craniovertebral instability seems to be a reasonable strategy of surgical treatment. Although direct screw implantation into the lateral mass of atlas in the presence of occipitization of the atlas is a relatively difficult surgical procedure, we found that the screw purchase and stability achieved was much stronger than screw implantation or wire fixation in the occipital squama which is relatively a thin shell of bone. Direct application of screws to atlas and axis, thus utilizing the firm purchase in their thick and large corticocancellous lateral mass, provided a biomechanically strong fixation of the region. Our observation is that although technically difficult and anatomically precise, lateral mass plate and screw fixation is an ideal method of fixation of atlantoaxial dislocation in cases with occipitized atlas. It may be the only option in cases where posterior foramen magnum decompression is necessary or has already been performed.\textsuperscript{[21]}

**Transoral decompression and fixation:** A number of craniovertebral anomalies that required transoral surgical decompression are all currently treated in our department by the technique of atlantoaxial joint distraction. The remarkable clinical result that we have achieved with the distraction technique makes us feel that transoral surgical decompression could become an operation of historical interest only.

Transoral surgery for various lesions in the craniocervical region has been used for a long time. Instability of the craniocervical region and development of kyphus after odontoidectomy in an otherwise stable joint has been reported. Most authors describe posterior fixation of the atlantoaxial region after transoral odontoidectomy. Satisfactory results have been obtained by such twin procedures.

We had described the feasibility of anterior transoral plate and screw fixation of the craniocervical junction after transoral decompression in the year 1994.\textsuperscript{[22]} Although the procedure of transoral plate and screw fixation did not become extremely popular due to the potential risk of infection, there are occasional indications for such a procedure and some groups recommend the use of transoral implants. The highly vascular nasopharyngeal mucosa has excellent healing potential and can accept grafts. Although the anterior transoral plate fixation is technically challenging and fraught with the dangers of infection, it can be useful in a rare case, particularly in a well-nourished patient with good oral hygiene. The fixation appears to be good, with clivus and the cervical body forming a strong ground for screws. The second operation of posterior fixation can be avoided. It has been suggested that our transoral plate fixation system can be performed by a transsurgical or retropharyngeal surgery, circumventing the risk of infection.

**References**