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Original Article

Post-operative Morbidity after C2 Nerve Root Sacrifice during C1-2 Fixation in Pediatric Atlantoaxial Instability

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BACKGROUND: The presence of the C2 root ganglion with its surrounding venous plexus at the region of C1-2 joint makes surgical dissection technically demanding. Although it may result in occipital sensory deficits, C2 ganglion neurectomy improves visualization of the C1-C2 joint and allows for control of venous bleeding. Therefore, the clinical benefits of C2 ganglion neurectomy in C1-C2 fixation are still controversial.

OBJECTIVE: To investigate the clinical outcome of pediatric atlantoaxial instrumentation using the Goel-Harms technique after C2 root sacrifice.

PATIENTS AND METHODS: This is a prospective cohort study including 20 pediatric patients with atlantoaxial instability. Outcome was assessed by visual analog scale (VAS) score for neck pain, in addition to sensory examination of the occipital region for assessment of any sensory deficits.

RESULTS: The mean operative time was 145 ± 16.38 minutes. The mean estimated blood loss was 211.5 ± 13.5 ml.
Thirteen cases (65%) had early occipital anaesthesia, while 7 cases (35%) had occipital dysthesia.
After 2 years of follow-up, resolution of sensory deficits was the rule in most cases, while only 4 cases had mild occipital dysthesia.
The VAS score for neck pain improved significantly from a mean of 6.15 ± 0 .93 preoperative to a mean of 1.30 ± 0.66 at 2 years postoperative ($p<0.001$).

CONCLUSION: C2 root sectioning during C1-2 fixation leads to better visualization of the C1 lateral mass besides less blood loss, but leads to early postoperative sensory deficits. However, complete resolution of these deficits was the rule in most cases after 2 years of follow-up.

KEYWORDS: Atlantoaxial instability, C1-2 fixation, C2 sacrifice.

INTRODUCTION

Goel-Harms technique for C1-C2 instrumentation is a relatively recent surgical procedure that could treat diverse causes of atlantoaxial instability. This technique involves atlantoaxial arthrodesis via insertion of polyaxial screws and rods. The screws are inserted into the C1 lateral masses and the C2 pedicles.[1,2] However, surgical dissection at the region of C1-C2 joint is technically demanding and usually faced with excessive bleeding. This is due to the presence of the C2 root ganglion with its surrounding venous plexus at the region of C1-C2 joint.[3,4] C2 ganglion neurectomy provides good visualization of C1-C2 joint and hence, panoramic visualization of the C1 lateral mass. Also, resection of C2 ganglion is supposed to be associated with less blood loss and operation time.[4,5] Occipital hyposthesia, numbness and neuropathic ulcers are potential complications of C2 ganglion neurectomy.[4-6] Yet, preservation of C2 nerve root per se has been associated with occipital dysthesia and numbness due to C2 nerve entrapment.[4,7,8] Therefore, clinical benefits of C2 ganglion neurectomy in C1-C2 fixation is still controversial.[6,7,9,10] This study aimed to investigate the clinical outcome of atlantoaxial instrumentation using Goel-Harms technique after C2 root sacrifice.

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PATIENTS AND METHODS

This is a prospective cohort study that was conducted at the Neurosurgery Department of Alexandria University Hospitals after obtaining approval from the local ethical committee and Institutional Review Board (IRB) of Alexandria University (Serial number 0306076). Data was collected from the database of 20 pediatric patients who suffered from atlantoaxial instability. Patients were operated for atlantoaxial fixation using Goel-Harms technique at a single institution and followed up for at least 24 months. The aim was to evaluate the pros and cons of pediatric C1-2 screw-rod fixation after C2 root sacrifice. There were 11 females and 9 males. The mean age at the time of the operation was 14.2 ± 1.99 years with a range from 11 – 17 years. The indications for atlantoaxial fixation included 6 cases of os odontoideum, 6 cases of dens fracture, 3 cases of Down syndrome, 3 cases of atlantoaxial rotatory fixation (AARF), and 2 cases of basilar invagination **(Figs. 1, 2).**

Os odontoideum is a smooth cortical bony ossicle

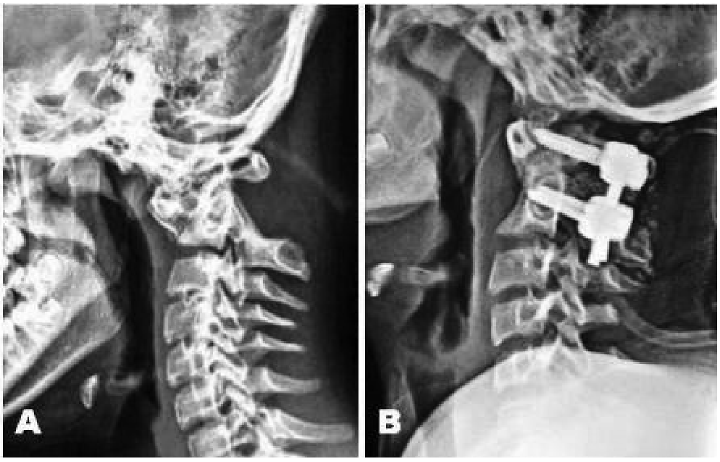

Fig 1: (A) Plain X-ray film of the cervical spine (lateral view) in a 13-year-old female Down patient showing atlantoaxial instability (B). Immediate postoperative X-ray film (lateral view) showing sound fixation and good reduction of C1-2 (B).

Fig 2: (A) Plain X-ray film of the cervical spine (lateral view) in a 15-year-old female patient showing atlantoaxial instability due to atlantoaxial rotatory fixation. (B) Cervical axial computerized (CT) scan of the same patient showing the rotatory fixation of C1 over C2. (C) Immediate postoperative sagittal CT scan of the cervical spine showed good alignment of C1 over C2. (D) Immediate postoperative axial CT scan of the cervical spine showed reduction of the rotatory fixation with the C1-2 screws.

representing a rudimentary odontoid process. It is not continuous with the rest of C2 body,[11] all the 6 cases of os odontoideum in the study were associated with atlantoaxial instability. The 6 cases of dens fracture were type 2 fractures. The 3 cases with Down syndrome were associated with atlantoaxial instability due to the excessive laxity of the ligaments including the transverse ligament. The 3 cases of AARF were associated with trauma. The 2 cases of basilar invagination were associated with Chiari malformation and scoliosis.

Before the operation, written informed consent was obtained from each patient's guardian to be included in the research study, and each patient was informed about the purpose of the study, the details of the procedure, the benefits, and the drawbacks of the technique. All patients were operated for atlantoaxial fixation by senior spine surgeons. Polyaxial screws and rods were inserted into the C1 lateral masses and the C2 pedicles. Plain X rays and computerized tomography (CT) scans were used for postoperative radiological evaluation. For a minimum of 48 months, the patients were evaluated through clinical and radiological follow-up.

Outcome measures: The VAS score for neck pain and early and late (2yrs) postoperative sensory deficits were used for clinical evaluation of the involved patients. In addition to CT scanning for a more thorough examination, radiological evaluation was obtained using plain-X rays of the instrumented segments in both anteroposterior and lateral views.

Data was fed to the computer and IBM statistical package for social sciences software version 20.0 was used for analysis (Armonk, NY: IBM Corp). Numerical and percentage representations were used for categorical data. For continuous data, the Shapiro-Wilk test was used to check for normality. Quantitative data were expressed as range (minimum and maximum), mean, standard deviation and median. When comparing two periods of non-normally distributed quantitative variables, the Wilcoxon signed ranks test was employed. Significance of the obtained results was judged at the 5% level.

Surgical technique

After general anesthesia and careful prone positioning, exposure of the occiput, C1, C2 and C3 was adopted via subperiosteal dissection of the attached occipital and suboccipital muscles. Exposure of the region of C1-2 facet joint was done via rostral subperiosteal dissection along the pedicle of C2 till reaching the C2 root ganglion which is located just posterior to the C1-2 joint. Wide exposure followed by sharp sectioning of C2 root ganglion bilaterally permits adequate visualization of the joint capsule and facilitates control of bleeding points from the peri-radicular venous plexus. It is important to avoid removing the ganglion too close to the dural ring to prevent postoperative cerebrospinal fluid leak. A proximal stump (3-4mm) of the preganglionic segment of C2 is left attached to the dural ring and could be coagulated. After adequate haemostasis, the joint capsule should be cut in a sharp way to allow for adequate decortication of the articular surface. The screws are then inserted into the lateral mass of the atlas and the pedicle of the axis. The preferred insertion point for the lateral mass screw of the atlas is at the center of the posterior surface of the lateral mass just above the articular surface. The screw insertion begins with drilling of the entry point by high speed drill. The screw is then directed about 15 degrees medially and superiorly guided by the C arm. Regarding the C2 pedicle screw, the medial surface of the pedicle should be identified and palpated before screw insertion. The preferred entry point is the superior and medial part of the axis pars. After drilling of the entry point, the screw is inserted and is directed about 25 degrees medially, and 15 degrees superiorly towards the superior part of the axis body. The screws are usually 3 mm in diameter, and 22-28 mm in length. Preparation of the surgical bed for bony fusion is done with bony decortication and removal of any intervening soft tissue. Rods are then inserted over the polyaxial screws to achieve reduction and stability **(Fig. 3)**. Autogenous bone graft harvested from the iliac crest is then inserted to enhance fusion. Postoperative external neck support is needed for at least 3 months.

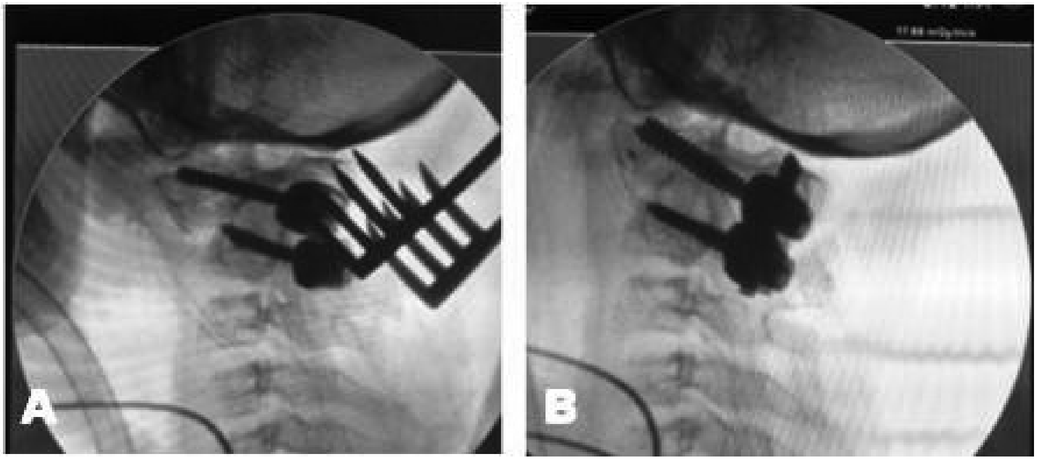

Fig 3: Intraoperative X-ray image of the craniocervical junction in a pediatric case with atlantoaxial subluxation showing (A) The anterior subluxation of C1 over C2 with the screws inserted and (B) reduction of C1 over C2 after rod insertion.

RESULTS

The study included 20 pediatric patients with atlantoaxial instability. There were 9 males and 11 females. The mean age at the time of the operation was 14.2 ± 1.99 years with a range from 11 to 17 years. Neck pain was a universal clinical finding in all cases (100%). Cervical myelopathy was the second most common presentation in 9 cases (45%). The third most common presentation was restricted neck movements in 6 cases (30%). Torticollis was found in 3 cases (15%), and scoliosis in 2 cases (10%). The mean operative time was 145 ± 16.38 minutes with a range from 120 – 180 minutes. The mean estimated blood loss was 211.5 ± 13.5 ml with a range from 190 – 240 ml.

The mean follow up was 30.4 ± 5.86 months. The most common early postoperative complication was occipital anaesthesia in 13 cases (65%) followed by occipital dysthesia in 7 cases (35%). After 2 years of follow up, 4 cases (20%) had mild occipital dysthesia, while the remaining 16 (80%) cases were free from any sensory manifestations.

The VAS score for neck pain has improved significantly from a mean of 6.15 ± 0.93, preoperative, to a mean of 1.30 ± 0.66 at 2 years postoperative ($p<0.001$) **(Tables 1-3).**

Table 1: Clinical and demographic characteristics of study patients and clinical outcome

	Age	Sex	Pathology	Clinical presentation	Operative time (minutes)	Blood loss (cc)	Early Postoperative complications	2 yrs Postoperative outcome	VAS score (pre-2 yr post)	FU* (months)
1	13	F	Down	Neck pain, Myelopathy	120	230	Occipital dysthesia	resolved	6-2	24
2	15	M	AARF*	Neck pain, Torticollis	130	220	Occipital dysthesia	resolved	7-1	36
3	17	F	BI*	Neck pain, Scoliosis	150	230	Occipital anasthesia	Occipital dysthesia	5-2	48
4	16	M	AARF	Neck pain, Torticollis	160	210	Occipital anasthesia	resolved	6-2	40
5	11	F	Os*	Neck pain, Myelopathy	150	190	Occipital anasthesia	resolved	5-1	25
6	13	F	Os	Neck pain, Myelopathy	140	200	Occipital anasthesia	resolved	5-2	25
7	14	M	Os	Neck pain, Myelopthy	150	220	Occipital anasthesia	Occipital dysthesia	5-1	28
8	12	F	AARF	Neck pain, Torticollis	130	220	Occipital dysthesia	resolved	7-2	30
9	11	F	Os	Neck pain, Myelopathy	130	200	Occipital anasthesia	resolved	6-1	24
10	13	M	Os	Neck pain, Myelopathy	150	220	Occipital anasthesia	resolved	6-1	26
11	14	M	BI	Neck pain, Scoliosis	130	210	Occipital anasthesia	Occipital dysthesia	5-1	28
12	16	M	Dens fracture	Neck pain, Restricted neck movements	150	200	Occipital anasthesia	resolved	7-0	28
13	14	F	Os	Neck pain, Myelopathy	150	190	Occipital anasthesia	resolved	6-1	27
14	15	F	Dens fracture	Neck pain, Restricted neck movements	140	210	Occipital anasthesia	resolved	8-1	30
15	14	F	Down	Neck pain, Myelopathy	180	210	Occipital dysthesia	resolved	5-3	32
16	16	M	Dens fracture	Neck pain, Restricted neck movements	160	220	Occipital dysthesia	resolved	7-1	30
17	15	F	Dens fracture	Neck pain, Restricted neck movements	130	210	Occipital anasthesia	Occipital dysthesia	7-1	34
18	11	M	Down	Neck pain, Myelopathy	130	200	Occipital dysthesia	resolved	6-1	32
19	17	M	Dens fracture	Neck pain, Restricted neck movements	140	200	Occipital dysthesia	Resolved	7-1	33
20	17	F	Dens fracture	Neck pain, Restricted neck movements	180	240	Occipital anasthesia	resolved	7-1	28

FU; follow up,,, AARF; atlantoaxial rotatory fixation,,, BI;basilar invagination,,, Os; os odontoideum.

Table 2: Distribution of the studied cases according to different parameters

	No. (%)
Age (years)	
Mean ± SD.	14.2 ± 1.99
Median (Minimum – Maximum)	14 (11 – 17)
Sex	
Male	9 (45%)
Female	11 (55%)
Pathology	
Atlantoaxial rotatory fixation	3 (15%)
Os odontoideum	6 (30%)
Down syndrome	3 (15%)
Dens fracture	6 (30%)
Basilar invagination	2 (10%)
Clinical presentation	
Neck pain, Myelopathy	9 (45%)
Neck pain, Torticollis	3 (15%)
Neck pain, Scoliosis	2 (10%)
Neck pain, Restricted neck movements	6 (30%)
Operative time (minutes)	
Mean ± SD.	145 ± 16.38
Median (Minimum – Maximum)	145 (120 – 180)
Blood loss (cc)	
Mean ± SD.	211.5 ± 13.5
Median (Minimum – Maximum)	210 (190 – 240)
Early Postoperative complications	
Occipital dysthesia	7 (35%)
Occipital anaesthesia	13 (65%)
Late Postoperative outcome	
Occipital dysthesia	4 (20%)
Resolved	16 (80%)
Follow-up (months)	
Mean ± SD.	30.4 ± 5.86
Median (Minimum – Maximum)	29 (24 – 48)

SD: Standard deviation.

Table 3: Comparison between preoperative and 2 years postoperative VAS score for neck pain

VAS score	Pre-operative	2 years post-operative	Z	p
Mean ± SD.	6.15 ± 0 .93	1.30 ± 0.66	3.941*	<0.001*
Median (Min. – Max.)	7 (5 – 8)	1 (0 – 3)		

SD: Standard deviation. Z: Wilcoxon signed ranks test.
p: p value for comparing between Preoperative and 2 years post operative score.

*: Statistically significant at $p \leq 0.05$.

DISCUSSION

The pediatric atlantoaxial region has some characteristics that make it more vulnerable to injury. These characteristics include less mature bone, laxity of the ligaments, horizontal facet joints orientation, and large head to body mass ratio with higher torque forces. There are many known causes of pediatric atlantoaxial instability, namely Down syndrome (DS), dens fracture, atlantoaxial rotatory fixation (AARF) and Os odontoideum. The present study had a total of 20 cases of pediatric atlantoaxial instability; 6 cases with type II dens fracture, 6 cases with os odontoideum, 3 cases with AARF, 3 cases with Down syndrome, and 2 cases of basilar invagination. As regards the clinical presentation; neck pain was a universal finding in all cases, cervical myelopathy was the main clinical manifestation in the 3 cases of Down syndrome and the 6 cases of Os odontoideum, restricted neck movements were encountered in the 6 cases with dens fracture, torticollis was the main presenting complain in the 3 cases of AARF, while the 2 cases of basilar invagination were associated with scoliosis. Treatment of pediatric atlantoaxial instability is challenging. Decompressing the neural components is the primary aim of surgical treatment, in addition to establishing a stable sound construct.[12-17]

The purpose of this study was to evaluate the pros and cons of pediatric C1-2 screw-rod fixation after C2 nerve root sacrifice. Although there are many surgical options available for treatment of pediatric atlantoaxial instability, but the authors prefer the screw-rod technique originally described by Goel-Harms due to its minimal complications and higher fusion rates when compared to other techniques.[1,2,17] Several authors emphasized the advantages of C2 root sectioning during surgical exposure of the C1 lateral mass, including improved visualization of the C1 lateral mass besides less blood loss and shorter operative time.[5-7] In a recent article, Abou Madawi et al. had examined the feasibility and safety of C1-C2 fusion in the treatment of pediatric atlantoaxial instability. They included 25 pediatric patients and reported less operative time and blood loss in patients who underwent C2 root sectioning although not statistically significant. The mean operative time for patients undergoing C2 neurotomy was 118.6 ± 9.2 minutes, while for other patients it was 136.8 ± 11 minutes. In addition, in patients undergoing C2 neurotomy, the estimated blood loss was 222.7 ± 81.7 ml, whereas in other patients, it was 345 ± 138.3 ml.[18] In the current study, the mean operative time was 145 ± 16.38 minutes, and the mean estimated blood loss was 211.5 ± 13.5 ml. However, this approach is technically challenging and may lead to troublesome sensory dysthesia in the occipital region. Occipital neuropathic ulcers have been associated with C2 root sectioning and have been reported. in cases with congenital atlantoaxial dislocations,[6,19,20] Also, occipital numbness and anaesthesia have been reported by some authors.[10,21,22] Rezvani et al. reported occipital anaesthesia as the most common postoperative complication following C2 sacrifice for C1-2 fixation in 81.5% of cases. They also reported occipital parasthesia in 63% of cases and mild pain in 66.7% of cases. Most of these sensory deficits resolved after the first 6 months of follow up.[23] In this study, all the included cases suffered from early sensory deficits in the form of occipital anaesthesia (13 cases) and occipital dysthesia (7 cases). These senosory deficits were often obscured by postoperative neck pain. Complete resolution of the sensory deficits was the rule in most patients (16 cases) after 2 years of follow up. The authors reported neither cases of neuropathic Occipital ulcers, nor cases of occipital neuralgia. In contrast, some authors have reported occipital neuralgia after C2 root preservation in about 5-35% of cases.[4,7] Serial clinical and radiological follow up showed 100% fusion rate with significant improvement of neck pain; the mean preoperative VAS score for neck pain was 6.15 ± 0 .93 and was decreased to a mean score of 1.30 ± 0.66 at 2 years postoperative (p <0.001).

CONCLUSION

While C2 root sectioning during surgical exposure of the C1 lateral mass has many benefits, including reduced blood loss and improved visualization of the C1 lateral mass, it also has some drawbacks in terms of early postoperative sensory deficits, mostly in the form of occipital anaesthesia, and to a lesser extent troublesome dysthesia. Nevertheless, following surgery, these deficits were frequently masked by neck pain, and after two years of follow-up, most cases showed complete resolution.

List of Abbreviation

AARF: Atlantoaxial rotatory fixation.
BI: Basilar invagination.
CT: Computerized tomography.
BI: Basilar invagination.
DS: Downs syndrome.
FU: Follow up.
IRP: Institutional review board.
Os: Os odontoideum.
SD: Standard deviation.
SPSS: Statistical packages for social sciences.
VAS score: Visual analogue score.

Disclosure

The authors report no conflict of interest in the materials or methods used in this study or the findings specified in this paper.

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