Ann Natl Acad Med Sci (India), **52(2):** 131-138, 2016

High cervical myelopathy due to bony craniovertebral junction anomalies (atlantoaxial dislocation) in pediatric population- clinical scoring system

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SUMMARY

Bony craniovertebral junction anomalies are rare anomalies to cause high cervical myelopathy. Atlantoaxial dislocation (congenital) is one of the commonest bony anomaly in children presenting with high cervical compression. It is relatively common in India with an incidence of 5-8 / 1000. When the distance of atlas (anterior arch) is more than 3mm (4 mm children) from odontoid process, it is called as Atlantoaxial dislocation (AAD) resulting into bony compression of high cervical cord. The patients may present with quadriparesis, sensory impairment in all limbs along with lower cranial nerve involvement. Because of lower medullary involvement the respiratory compromises are also frequent, posing a threat to life. Complex anatomy of foramen magnum, plethora of clinical conditions and atypical surgical approaches are responsible for poor outcome in these children. A new clinical scoring system for myelopathy was evolved in order to have an objective and precise grading of these cases preoperatively and postoperatively. The need of precise scoring system was felt to have reproducibility and easy applicability in children of craniovertebral junction anomalies in order to fetch even minimal improvement or deterioration following complex surgery. Motor functions, gait, sensory, sphincteric, respiratory function & spasticity were the parameters included in study of scoring system. This study was done in 177 operated cases of AAD (67 patients, below 14 years of age included for statistical analysis). Their detailed clinical & radiological evaluation was done preoperatively & postoperatively. The Kumar & Kalra high cervical myelopathy grading system was thus, introduced in literature. System was easy to use, interpret and was more sensitive

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DR. S. JANAKI MEMORIAL ORATION delivered during NAMSCON 2015 at the All-India Institute of Medical Sciences, Patna.

to the changes in neurological status. It helped neurosurgeons and neurologists globally to evaluate and prognosticate the cases of Atlantoaxial dislocation.

Keywords : Pediatric, atlantoaxial dislocation, CV Junction anomalies, high cervical myelopathy.

The craniovertebral junction (CVJ) is a complex transition zone between the cranial cavity and the rostral spinal column. Various conditions affect and destabilize this region; of these conditions, congenital atlantoaxial dislocation (AAD) is the most common anomaly (1, 2). The management of congenital CVJ anomalies is complex due to the relative difficulty in accessing the region, the critical relationship of neurovascular structures, and the intricate biomechanical issues involved, which revolve primarily around the attainment of neural decompression and stabilization (3). The difficulty accessing the CVJ relates primarily to ventrally compressing pathologies in which the transoral procedure is required to take out the offending bony element. In comparison, posterior access to the CVJ does not pose much of a problem but has threat of neural injury.

The most common age group affected by congenital AAD is the second and third decade; however, the incidence in the paediatric population is not uncommon. A wide variety of congenital anomalies of the CVJ occur due to an insult of bony and soft tissue elements, between the fourth and seventh week of intrauterine life (4). The clinical manifestations are most often delayed into the second and third decade because they are usually subtle and easily missed in children unless looked for specifically (1, 5). Moreover, this disease process is slowly progressive and manifests due to chronic compression of the cervicomedullary junction; it is usually brought about by triggering events such as trauma (1, 5, 6). The criteria for diagnosis of AAD differ between children and adults. The normal atlas to dens interval is greater in children compared with adults; thus, AAD is diagnosed on the basis of atlantodental distance greater than 4 mm in patients younger than 9 years of age and greater than 3 mm for those 9 years of age or older (6). The management of these patients becomes even more difficult as a result of the general issues of managing children, which add to the typical issues of managing the complicated anomaly itself. There has been a progressive change in the approaches and techniques used for management (7). The outcome analysis performed to assess the surgical result has relied on Di Lorenzo's grading (DLG) system, which is a four-grade system based on functional dependence (Table 1) (3, 5, 8). This has been the most widely used and accepted grading system to

assess AAD patients undergoing treatment. The grading system has stood the test of time and has excellent reproducibility and inter observer reliability (7). However, although the DLG grading system is universally acceptable in adults, it may not be as readily applicable and useful in children. Children are unable to quantify their functional status and are otherwise dependent for their daily activities. Hence, it is difficult to access their functional disabilities caused by the disease process. Categorization according to disabilities as is done in the DLG may not be precise in this age group and lacks interobserver reliability and reproducibility.

Moreover, grading the patients into just four groups does not allow for the observation of subtle changes in neurological status. There is a large subset of patients who remain in the same grade even though they have subjective improvement or deterioration in their neurological status. It may be opined that a comprehensive and precise neurological examination may be more valuable in accessing the response to treatment. If a scoring system is developed that relies on these factors, it may be better reflective of progression of the disease course. Such a score should be detailed enough to recognize subtle neurological status changes but simple enough to be performed routinely. The purpose of the present study was to formulate a new scoring system based on comprehensive neurological examination and to assess its applicability in children compared with the DLG.

Table 1 : Di Lorenzo's Grading system

Grade 1	Independent, without deficits except hyperreflexia or neck pain. (Neurologically intact)
Grade 2	Independent for daily activities but having minor deficits (minor disability)
Grade 3	Partly dependent on others for their daily needs (moderate disability)
Grade 4	Totally dependent on others for daily needs (severe disability)

A study was done at SGPGIMS, Lucknow to introduce new scoring system; this was based on clinico-radiological evaluation of 177 cases operated over period of 14 years by single surgeon (RK). Statistical analysis was performed on children with age less than 14 years (n=67). The diagnosis was based on a minimum atlantodental interval of 4 mm. The patients who had associated Chiari- I malformation, AAD and associated genetic syndromes , or follow up of Less than 12 months were excluded. The assessment of patients was done preoperatively and postoperatively & follow up. The following points affecting the outcome were taken into consideration to fetch a new and precise scoring system

for better evaluation of children suffering from congenital AAD (Table 2). There were 36 males and 31 females, with a mean age 9.36±3.51 years (range 2-14 years). The results were compared with Di Lorenzo grading system.

Table 2 : Kumar & Kalra scoring system (K & K)

Parameter	**1**	**2**	**3**	**4**	**5**	**Total score**
Motor power	Contraction w/o movement or plegia	Movement with gravity	Movement against gravity	Movement against resistance	Normal power	5
Gait	Wheel chair bound or bed ridden	Restricted mobility despite aid	Mobility using aid	Slight disturbance, no aid required	Normal	5
Sensory involvement	Total loss of function	Restriction of function of daily living	Significant involvement (>25%) but no dysfunction of daily living	insignificant	No sensory loss	5
Sphincter involvement	Retention requiring indwelling catheter	Occasional CIC required with hesitancy	Hesitancy with residual urine not requiring catheter	Hesitancy but no residual urine	Normal	5
Spasticity	Affected part rigid in flexion or extension	Passive movement difficult	Passive movement easy	Slight increase in tone	Normal	5
Respiratory difficulty	Requires assisted respiration	Dyspnoea at rest	Dyspnoea on mild exertion	Dyspnoea on moderate exertion, unable to do active work	Normal	5

Disability Grading :

67 patients, treated surgically for AAD and having age less than 14 years, included in this study. They were assessed with the DLG and Kumar and Kalra (K&K) scores pre-and postoperatively (Table 3 and 4). The patients were divided into four grades according to the K&K scores, with grade 1 patients having score from 25 to 30, Grade 2 patients having score from 19 to 24, Grade 3 patients having score from 13 to 18 and Grade 4 patients having score from 6 to 12 points. Most of the patients had poor grades. Using the DLG scoring system, there were 26 (38.8%) patients in Grade 4 and seven (10.4%) patients in Grade 3 preoperatively. The maximum postoperative improvement was witnessed in the Grade 4 patients, with as many as 23 patients showing improvement from their preoperative grades. Overall, 47.8% of the patients showed improvement and 10.4% showed deterioration. There was a high percentage (41.8%) of patients who showed stabilization of their grades. When the K&K scoring system was used on these patients, most (65.7%) of the patients were rated as Grade 2 preoperatively. The number of patients (n=13) in preoperative Grade 1 was also high. Only three patients were rated as Grade 4 preoperatively, and none remained in this group postoperatively. Overall, improvement was observed in the majority (n = 55) of the patients; fewer patients had deterioration (n = 6). Thus, stabilization of neurological status was seen in 41.8% of patients using the DLG score and in only 8.9% of the patients using the K&K score. Although 47.8% of the patients showed improvement using the DLG scoring system, a very high percentage (82.1%) of patients showed improvement using the K&K score. Out of the 28 patients who had shown stabilization of their neurological status using the DLG score, 23 patients showed an improvement in scores when assessed by the K&K score, two patients showed the stabilization of their scores, and three had deteriorated from their preoperative scores. The minimum improvement was observed in those with symptoms of weakness (n = 49) and spasticity (n = 47), which were the earliest symptoms to improve. The symptoms most resistant to treatment were respiratory (n = 7) and sphincter symptoms (n = 6). To establish the objective criteria of reliability and reproducibility of the proposed score, we analyzed each patient's DLG and K&K scores preoperatively and compared them with their most recent follow-up scores and outcome. The statistical significance was ascertained by calculating p-value using logistic regression and by measuring the positive predictive values for each score. The results are listed in Table 5.

Table 3 : Outcome analysis based on Di Lorenzo's grading system

Grade	Pretreatment, n= 67 (100%)	Posttreatment n=67 (100%)	Improved (%)	Same (%)	Deteriorated (%)
I	10 (14.9)	17 (25.4)	0	8	2
II	24 (35.8)	19 (28.4)	8	14	2
III	7 (10.4)	25 (37.3)	1	3	3
IV	26 (38.8)	6 (9)	23	3	0
Total			32 (47.8)	28 (41.8)	7 (10.4)

Table 4 : Outcome analysis based on Kumar & Kalra score

Grade*	Pretreatment, n= 67 (100%)	Posttreatment n=67 (100%)	Improved (%)	Same (%)	Deteriorated (%)
I (25-30)	13 (19.4)	17 (25.4)	11	1	1
II (19-24)	44 (65.7)	45 (67.2)	34	5	5
III (13-18)	7 (10.4)	5 (7.5)	7	0	0
IV (6-12)	3 (4.5)	0	3	0	0
Total			55 (82.1)	6 (8.9)	6 (8.9)

*Numbers in parentheses in the first column represent the value of the total score using the Kumar and Kalra score.

Table 5: Predictability of score

Score used	Mean pretreatment	Mean follow up value	P value	Exp (B)	Predictive value (%)
DLG	2.73 $\pm$ 1.14	2.30$\pm$ 0.95	0.714	1.094	52.2
K&K	19.91$\pm$ 4	23.22$\pm$ 3.36	0.000	0.218	82.1

DLG- Di Lorenzo's grade; K&K- Kumar & Kalra score.

The incidence of AAD in pediatric age group is significant enough to deserve special and separate emphasis. Majority of children with AAD in our country reach neurosurgeons through referral system and thus usually present in poor grades. After surgery, it has been observed that although majority of them show subjective improvement, a large number stay in same grade as their pre operative grade when DLG scoring system is used for assessment (1, 5). Furthermore there are patients who deteriorate & may require surgery but stay in same grade. The DLG score used for adults is lacking in its reproducibility in children and does not truly reflect the gravity of disease process. Thus, it is not a very sensitive indicator of change in neurological status and cannot be reliable in children. The change in score and neurological status is better reflected by K&K scores rather than

DLG. Positive predictive value was better using K &K score , and outcome analysis using K&K score was statistically significant whereas the changes didn't bear statistically significant correlation using the DLG score (p=0.714). The advantages are the assessment and comparison of neurological status based on commonly occurring symptoms and signs, which are routinely assessed in neurological examination of such patients. The individual symptoms and signs can also be analysed and compared individually in terms of their severity and significance. Thus, both an overall status of function and individual symptom is known. The score is based on multitude of factors and hence is more sensitive. It is also more suited for statistical analysis. The score may very well be used for adult patients as the parameters used are common to adults also. Furthermore, after exclusion of respiratory parameters, it can also be used for myelopathy in other areas of spine.

To conclude, K & K scoring system is easy to use and interpret, and is more sensitive to the changes in neurological status of patients than the currently used DLG system (9).

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