Unusual cause of high cervical myelopathy-C1 arch stenosis

ABSTRACT
Introduction: High cervical myelopathy can be rarely caused by the developmental anomalies of atlas. Patients with C1 arch stenosis can present in early childhood or later in life. In symptomatic patients, posterior decompression at atlas is mandatory. We report the first clinical series of 20 patients of C1 arch stenosis in the English literature.

Materials and Methods: This is retrospective case series having a cohort of 20 patients with congenital C1 arch stenosis.

Results: There were 12 pediatric (age <18 years) and 8 adult patients. Mean age was 22.85 years. Syndromic association was seen in four patients. Following decompressive surgery, these patients noticed a symptomatic improvement.

Conclusions: Isolated C1 arch stenosis is a surgically curable rare cause of high cervical myelopathy and responds well to surgery.

Keywords: Anomalies of atlas, bifid posterior arch, C1 arch stenosis, high cervical myelopathy, Morquio’s syndrome

INTRODUCTION

Developmental anomalies of the anterior and posterior arch of atlas can be a rare cause of high cervical myelopathy. The most common anomaly is a failure of posterior midline fusion of the two hemi-arches (Currarino classification Type A).

It may be seen with certain conditions such as achondroplasia, Turner’s syndrome, and various storage disorders, e.g., mucopolysaccharidosis.

Patients with C1 arch stenosis can present in early childhood or later in life. Many times, the C1 arch stenosis is not diagnosed and may be missed and labeled as “no cause found.” Some of these patients may present later in life with degenerative cervical spine and managed accordingly even though the C1 shows stenosis.

Earlier, we had reported five children with C1 arch stenosis managed by senior author. Over the past 20 years, we managed a total of 20 cases with C1 arch stenosis. We report the first clinical series of 20 patients with C1 arch stenosis in the English literature.

MATERIALS AND METHODS

This present cohort includes 20 patients with congenital C1 arch stenosis. Patients with spinal canal compromise due to atlantoaxial dislocation at atlas were excluded. All patients were evaluated with dynamic radiography, computed tomography (CT) cervical spine, and magnetic resonance imaging (MRI) cervical spine. Following evaluation, all patients underwent posterior cervical decompression.

RESULTS

The study cohort includes 14 male and 6 female patients, aged 4–60 years (mean age 22.85 years). This study had 12 pediatric patients (age <18 years). Four patients had a syndromic association; two children and one adult with mucopolysaccharidosis Type 4 Morquio disease, adult, with neglected turricephaly. One adult patient had ankylosing spondylitis.

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The progressive spastic weakness of all four limbs (95%) was the most common clinical feature followed by a history of trivial fall (55%), neck pain (45%), dysmorphic features (20%) [Table 1]. Only two patients required surgery on emergent basis due to respiratory distress and recent neurological worsening. Clinical details, radiological features, and surgical findings are summarized in Table 2. At surgery inward, the inclination of the bifid C1 arch was seen.

**DISCUSSION**

Posterior arch of the atlas develops from the lateral dense zone of the first cervical sclerotome, while anterior arch of the atlas develops from hypochondral bow of the first cervical sclerotome ventral to the notochord.[5] Partial or complete agenesis of the posterior atlantal arch results from varying degrees of aplasia of the lateral sclerotome.[5]

The normal cervical vertebrae have a triangular spinal canal and are of a similar size to the vertebral body. The atlas has a ring-shaped spinal canal and lacks a vertebral body and a spinous process. Spinal canal diameter is maximum at the level of atlas and has the highest cord surface area-to-canal ratio. In C1 stenosis, canal becomes ellipsoid with reduced anteroposterior diameter, thus reducing the canal-to-cord ratio [Figure 1].

Stenosis at C1 can be congenital or acquired. Anomalies causing cervical myelopathy at the level of atlas are reported rarely in the literature and include clefts or aplasias of anterior and posterior arches of atlas, os odontoideum, ossification of the transverse atlantal ligament, and hypertrophy of the dens either alone or in combination.[6] Congenital stenosis of the cervical spinal canal most commonly occurs between C3 and C5 vertebrae.[3] Developmental stenosis of the cervical spine rarely occurs above C2.[7] The stenosis at the level of C1 may be due to excessive or disordered ossification. The posterior arch defects of C1 are uncommonly reported and the clinical significance is unknown. Generally asymptomatic, symptoms may be precipitated by head or neck trauma while playing. Eleven patients had a history of neck trauma secondary to fall or road traffic accident. Dynamic lateral cervical spine X-ray was done to look for instability. CT cervical spine is useful to demonstrate bony abnormality. MRI is useful in demonstrating site of compression and soft tissue anomalies. Table 3 highlights clinical and radiological details of previously reported cases of C1 arch stenosis in the literature.

The importance of congenital stenosis of the spinal canal was first recognized by Payne and Spillane, suggesting that the development of myelopathy may also be related to the initial size of canal.[14] If the spinal canal is small by nature relatively little encroachment by osteophyte, vertebral subluxation or hypertrophied ligament flavum can be tolerated without causing spinal cord compression. There have been a few reports as to the relative diameters of C1. According to Hinck et al., the sagittal diameter of the spinal canal at the level of C1 ranges between 15 mm and 20 mm.[15] A radiographic study of 300 normal Indians performed by Gupta et al. reported that the mean space available for the cord at C1 was 21.43 mm in male and 20.13 mm in female.[16] In our series, C1 canal diameter was 10.3 mm (range 0.5–16 mm).

The usually reported incidence of posterior arch anomalies of C1 is 4% and 0.1% for anterior arch.[17] The anomalies of posterior arch range from median clefts or hypoplasia. A failure of local chondrogenesis rather than subsequent ossification is thought as a cause for the development of

| Table 1: Common clinical features (n=20) |
|-----------------------------------------|
| **n (%)**                               |
| Dysmorphic facies                      | 4 (20) |
| Syndromic                              | 4 (20) |
| Trauma                                 | 11 (55) |
| Neck pain                              | 9 (45) |
| Neck tilt                              | 5 (25) |
| Respiratory difficulty                 | 2 (10) |
| Spasticity and/or weakness             | 19 (95) |

Figure 1: C1: (a) Normal ring-shaped atlas lacking vertebral body having maximum canal diameter. (b) In C1 stenosis: canal is ellipsoid with reduced anteroposterior diameter, thus reducing the canal-to-cord ratio. (c) Bifid posterior arch with inward inclination
Table 2: Clinical details, radiological features, and management

| Age | Sex | Clinical features                                                                 | Radiology                                      | Canal diameter (mm) | Surgery                              | Follow-up                                                                                     |
|-----|-----|-----------------------------------------------------------------------------------|-----------------------------------------------|---------------------|--------------------------------------|-----------------------------------------------------------------------------------------------|
|     |     | *(A) Pediatric nonsyndromic patients with stenosis at C1*                           |                                               |                     |                                      |                                                                                               |
| 1   | 10  | Male                                                                               | Frequent falls, clumsiness of movements       | Bifid posterior arch Klippel–Feil | 14                                   | Elective C1 arch excision                                                                    | 50 months: Spasticity reduced with reduced falls                                              |
|     |     |                                                                                   | dysmorphic facies. Torticollis, spastic       |                     |                                      |                                                                                               |
|     |     |                                                                                   | quadriaparesis with posterior column           |                     |                                      |                                                                                               |
|     |     |                                                                                   | impairment                                    |                     |                                      |                                                                                               |
| 2   | 6   | Male                                                                               | Neck pain following a fall, Spastic           | Bifid anterior and posterior arch, canal dia at C1 | 16                                   | Elective C1 arch excision                                                                    | 48 month: Spasticity reduced. Improved gait                                                  |
|     |     |                                                                                   | quadriaparesis with posterior column           |                     |                                      |                                                                                               |
|     |     |                                                                                   | impairment                                    |                     |                                      |                                                                                               |
| 3   | 15  | Male                                                                               | Neck tilt, neck pain, and paresthesia         | C1 arch stenosis with | 11                                   | Foramen magnum decompression with C1 arch excision                                           | At 1 month: Reduced paresthesia and neck pain                                                |
|     |     |                                                                                   | C4–C5 Klippel–Feil                            |                     |                                      |                                                                                               |
| 4   | 18  | Male                                                                               | Neck pain and neck tilt                       | C1 arch stenosis     | 11                                   | C1 posterior arch excision                                                                    | At 2 years, reduced neck pain                                                               |
|     |     |                                                                                   |                                               |                     |                                      |                                                                                               |
| 5   | 9   | Male                                                                               | Neck pain, quadriaparesis Developmental delay  | Posterior and anterior cleft of C1 arch in midline, C1 canal stenosis with cord myelomalacia, C2–C3 Klippel–Feil. [Figure 2] | 9                                   | C1 posterior arch excision and release of dural band                        | Follow-up at 8 months improved weakness (5/5)                                              |
|     |     |                                                                                   |                                               |                     |                                      |                                                                                               |
| 6   | 10  | Female                                                                             | Spastic quadriaparesis, not able to stand     | Cervical canal stenosis with cord compression | 8                                   | Excision of Posterior C1 arch                                                             | 3 months stand up with support                                                              |
|     |     |                                                                                   | without support and walk, delayed development |                                               |                                      |                                                                                               |
| 7   | 6   | Male                                                                               | Quadriplegia with respiratory distress         | Bifid anterior and posterior arch             | 16                                   | Emergency excision of C1 posterior arch. Postoperatively ventilation for 1 month. Tracheostomy | Off ventilator at discharge. Power 3/5.12 months walking with support. Spasticity minimally better |
|     |     |                                                                                   | following trivial fall. Frequent fall in past  |                                               |                                      |                                                                                               |
| 8   | 18  | Female                                                                             | Pain in the neck, progressive weakness in the  | Platybasia with C1 arch stenosis with small posterior fossa | 5.5                                 | C1 arch excision and foramen magnum decompression | Follow-up at 1 month walking independently                                                   |
|     |     |                                                                                   | left upper and lower limbs, walking with      |                                               |                                      |                                                                                               |
|     |     |                                                                                   | support, dysmorphic features                  |                                               |                                      |                                                                                               |
| 9   | 8   | Male                                                                               | Dysmorphic facies, torticollis since birth.   | Klippel-Feil, Bifid posterior arch at C1, C1 and hydromyelia extending up to C5 | 15                                   | Elective C1 arch excision                                                                    | 12 months no change in torticollis. Improved postcolumn and spasticity                      |
|     |     |                                                                                   | Frequent falls. Spastic quadriaparesis        |                                               |                                      |                                                                                               |
| 10  | 11  | Male                                                                               | Frequent falls, dysmorphic facies, and        | Bifid posterior arch                          | 16                                   | Emergency C1 arch excision. Postoperatively ventilator support | Discharge after closure of Tracheostomy. Slow improvement in spasticity. Follow-up 10 months |
|     |     |                                                                                   | torticollis spastic quadriplegia with         |                                               |                                      |                                                                                               |
|     |     |                                                                                   | respiratory distress                          |                                               |                                      |                                                                                               |
|     |     | *(B) Pediatric syndromic patients with stenosis at C1*                             |                                               |                     |                                      |                                                                                               |
| 11  | 4   | Female                                                                             | Trivial trauma, quadriaparesis, neck          | Incomplete fusion of anterior and posterior arch of atlas. [Figure 3] | 5                                   | C1 posterior arch excision                                                                    | 15 months walking independently                                                             |
|     |     |                                                                                   | deformity K/c/o/Morquio syndrome               |                                               |                                      |                                                                                               |

Contd...
Isolated C1 arch stenosis

Nehete, et al. showed the inward mobility of the posterior fragment during extension of the cervical spine in two patients as a possible reason for a patient with trivial trauma. The presence of connective tissue bridging the bony defect is supported by autopsies and intraoperative findings. Five different types of C1 arch deficiencies are described by Currarino et al. In this series, one patient had absent posterior arch (Currarino classification Type E). In this patient, fibrous band was seen compressing the cord. In two

| Age | Sex | Clinical features                                                                 | Radiology                                                                 | Canal diameter (mm) | Surgery                                      | Follow-up                        |
|-----|-----|-----------------------------------------------------------------------------------|---------------------------------------------------------------------------|--------------------|----------------------------------------------|-----------------------------------|
| 12  | 5   | Fall from chairs f/b quadriaparesis, failure to gain height, pectus carinatum, enlarged wrist, short fingers and toes, corneal cloudiness. | Spinal canal stenosis with hypoplasia of odontoid, cord signal changes at C1 | 7                  | C1 arch excision with C1, C2, C3 fusion     | Improved weakness at 6 months    |

**(B) Pediatric syndromic patients with stenosis at C1**

| Age | Sex | Clinical features                                                                 | Radiology                                                                 | Canal diameter (mm) | Surgery                                      | Follow-up                        |
|-----|-----|-----------------------------------------------------------------------------------|---------------------------------------------------------------------------|--------------------|----------------------------------------------|-----------------------------------|
| 13  | 47  | Multiple episodes of trauma/fall, neck pain, weakness of right upper and lower limb, low hairline, high arch palate | Stenosis at C1 with cord signal changes. [Figure 4]                       | 11                 | C1 posterior arch excision                    | At 3 months reduced neck pain, able to write |
| 14  | 52  | History of fall, bilateral upper and lower limb weakness and paraesthesia         | C1 canal stenosis and cord signal changes                                  | 7                  | C1 posterior arch excision                    | Improvement at 18 months: Walking independently |
| 15  | 19  | Dull headache, neck pain, all four limb weakness, brisk tendon reflexes          | Completely absent C1 posterior arch                                       | 11                 | Excision of dural band between foramen magnum and C2 | At 12 months reduced neck and headache. Improved weakness |
| 16  | 60  | Neck pain, frequent falls, progressive spastic quadriaparesis. Operated elsewhere as C2–C7 decompressive laminectomy | Stenosis at C1                                                             | 14                 | Posterior C1 arch excision                    | At 12 months spasticity better    |
| 17  | 35  | Multiple episodes of fall. Spastic quadriaparesis. Predominant posterior column impairment | C1 arch stenosis with cord signal changes at C1 and C2                    | 5.5                | C1 posterior arch excision and C2 laminectomy | Follow-up at 12th month: Minimal improvement |
| 18  | 54  | Diagnosed case of ankylosing spondylitis, neck pain with restriction of neck movements, gradually stiffness and progressing weakness of all four limbs | C1 arch stenosis with cord compression with cord signal changes at that level, straightening of cervical spine with C1 arch stenosis and bamboo stick appearance of vertebral body | 5                  | C1 posterior arch excision and C2 laminectomy | Improve limb movements and stiffness at 1 month follow-up |

**(C) Adult nonsyndromic patients with stenosis at C1 with cervical spondylosis**

| Age | Sex | Clinical features                                                                 | Radiology                                                                 | Canal diameter (mm) | Surgery                                      | Follow-up                        |
|-----|-----|-----------------------------------------------------------------------------------|---------------------------------------------------------------------------|--------------------|----------------------------------------------|-----------------------------------|
| 19  | 50  | Neck pain, quadriaparesis, dysmorphic facies tower shape skull                     | C1 arch stenosis, C3–C4 Klippel–Feil                                      | 10                 | C1 arch excision                            | Lost follow-up                   |
| 20  | 20  | Known case of Morquio syndrome, progressive spastic weakness of lower limbs after a fall. bilateral corneal clouding, restricted mouth opening, short and restricted neck, and receding chin | Fused C2 and C3 spinous processes. Soft tissue mass compressing at CVJ     | 9                  | Foramen magnum decompression, C1 posterior arch and soft tissue excision | At 10 months: Asymptomatic        |

CVJ - Craniovertebral–vertebral junction; f/b - Followed by; K/c/o - Known case of
Table 3: Clinical, radiological features and management of reported cases of C1 arch stenosis in the literature

| Number | Case report                                                                 | Age | Sex | Clinical details                                                                 | Imaging                                                                 | Management                          | Follow-up                        |
|--------|------------------------------------------------------------------------------|-----|-----|---------------------------------------------------------------------------------|------------------------------------------------------------------------|-------------------------------------|-----------------------------------|
| 1      | Spinal canal stenosis at the level of atlas, Bhattacharjee et al. [7]         | 10  | Male | Progressively deteriorating quadriparesis and respiratory distress              | Ill-defined osseous bar compressing the canal at the level of bifid C posterior arch | Surgery                             | Improved at 1 year               |
| 2      | Complete absence of the posterior arch of C1: Case report, Khanna et al. [8]  | 46  | Male | Neck pain since childhood                                                       | Total absence of the posterior elements of C1 and severe central canal stenosis at C3–4 | C3–C4 ACDF and Miami J collar       | 6 months improved myelopathic features |
| 3      | Hypertrophic posterior arch of atlas causing cervical myelopathy, Kasliwal and Traynelis [6] | 26  | Female | Neck pain with tingling and numbness of long duration                       | Hyperostotic right posterior arch of C1 with spondylosis               | Excision of C1 arch                 | 3 months improved                |
| 4      | Cervical myelopathy caused by hypoplasia of the atlas: Two case reports and review of the literature, Phan et al. [9] | Two elderly | Male | Gradually developing cervical myelopathy during months to years                | Hypoplastic but complete posterior C1 arch associated with changes of spondylosis, producing severe spinal stenosis and spinal cord compression | Removal of C1 posterior arch       | Improved at follow-up             |
| 5      | Spinal stenosis at the level of atlas in a boy with down syndrome. A case report and literature review. Pascual-Gallego et al. [10] | 5   | Male | Myelopathic features                                                            | Compression at C1                                                        | Removal of posterior C1 arch        | Improved                          |
| 6      | Spinal canal stenosis at the level of the atlas: Case report. Tokiyoshi et al. [11] | 55  | Male | Gait disturbance and clumsiness in both hands                                   | Spinal canal stenosis, with the diameter of 8.0 mm at the level of the atlas | Laminctomy of the atlas combined with decompression of the lower posterior fossa of the foramen magnum and dural plasty | N/A                              |
| 7      | Congenital midline cleft of the posterior arch of atlas: A rare cause of symptomatic cervical canal stenosis. Connor et al. [12] | 8   | Male | Difficulty while walking and clumsiness                                         | Inturned bifid posterior hemi-arches of C1 causing cord compression     | Excision of C1 posterior arch       | Improved at 6 week                |
| 8      | Partial Aplasia of the posterior arch of the atlas with an isolated posterior arch remnant: Findings in three cases Sharma et al. [13] | 32  | Female | Episodic weakness and numbness of all four limbs after sustaining a minor head trauma | Partial aplasia of the posterior arch of the atlas with focal hyperintensity of the cord | Advised surgery but lost follow-up | Lost follow-up                   |
|        |                                                                              | 35  | Female | Neck pain and a tingling sensation and weakness in both upper limbs            | Aplasia of the posterior arch of the atlas with focal hyperintensity of cord | Observe                             | Status quo                       |
|        |                                                                              | 30  | Male   | Recurrent neck pain and stiffness                                              | Partial aplasia of the posterior arch                                 | Observe                             | Status quo                       |

N/A - Not available

patients, failure of posterior midline fusion of posterior C1 arch was seen (Type A).

C1 arch stenosis may occur as part of a syndrome. It may be associated with Arnold–Chiari malformation, Klippel–Feil syndrome, gonadal dysgenesis, Down’s syndrome, and Turner’s syndrome. Three patients had mucopolysaccharidosis, Marquio syndrome. In mucopolysaccharidoses, the cord at the C1–C2 level is at particular risk for stenosis, ventrally by localized thickening...
Nehete, et al.: Isolated C1 arch stenosis

Figure 2: Pediatric: Pediatric patient with C1 arch stenosis. (a) Midsagittal computed tomography showing absent posterior arch in midline. (b) Inturned posterior arch causing canal narrowing with absent arch in midline and anterior arch midline cleft. (c and d): No evidence of instability on Dynamic X-ray. (e) Preoperative magnetic resonance imaging showing cord thinning at C1 with cord signal changes. (f) Postoperative magnetic resonance imaging showing expansion of cord with cerebrospinal fluid around

Figure 3: Syndromic: Morquio syndrome patient (a and b) computed tomography showing absent anterior and posterior arch in midline. (c) Magnetic resonance imaging cord thinning due to posterior soft tissue at C1

Figure 4: Adult: Adult patient of cervical spondylosis with C1 stenosis. (a) Computed tomography spine showing anteriorly placed posterior arch of C1 with canal narrowing. (b) Magnetic resonance imaging showing canal narrowing at C1 with cord signal changes

of the peri-odontoid tissue and transverse atlantoaxial ligament and dorsally due to a short narrow C1 posterior arch or anterior translation of the posterior arch of C1.[22] C1 arch stenosis in Morquio syndrome is due to narrow posterior arch with odontoid hypoplasia, soft tissue mass, or incomplete fusion of C1 arch. These patients require a preoperative evaluation of the airway and of cardiac, respiratory, and neurological function. Anesthetic management must focus on protection of the airway without compromising the integrity of the spinal cord.[23]

In addition to C1 arch stenosis, craniosynostosis was also seen in one of our patients. We think that impaired skull base cartilaginous ossification mechanism may be responsible for C1 arch stenosis since craniovertebral–vertebral junction is also part of the central skull base. Six patients in this series had Klippel–Feil syndrome.

All patients were managed with C1 arch excision. Only two patients required emergent surgery due to associated recent neurological deterioration and respiratory distress. Both
these patients required ventilator support postoperatively and were gradually weaned off the ventilator. During surgery intubated, knob-like bifid posterior arch was seen compressing cord in most of the patients. One patient with deficient posterior arch, the fibrous band, was seen compressing cord. After excision of a posterior arch, fibrous band cord was lax and pulsatile. On follow-up, we have not found any instability in patients managed with C1 arch excision. One patient with Morquio syndrome with associated odontoid hypoplasia underwent fusion after excision of C1 arch excision.

Our series on C1 arch stenosis is largest in the published literature till date and complements the cohort with different age groups. Awareness of C1 arch stenosis will improve with the published literature. Previously, asymptomatic C1 arch stenosis can present in the later life and needs to be managed with decompression since surgical treatment may prevent neurological deterioration.

CONCLUSIONS

Isolated C1 arch stenosis is a surgically curable rare cause of high cervical stenosis and responds well to surgery. It can be managed with C1 arch excision alone without subsequent development of instability. Unless surgically treated, minor trauma may cause the serious neurologic deficit.

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Informed consent

Informed consent was obtained from all individual participants included in the study.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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