SUBAXIAL DEFORMITIES OF THE CERVICAL SPINE

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ABSTRACT :

Several deformities of the cervical spine causes disability. These include a variety of degenerative, inflammatory, infectious, neoplastic, congenital, and traumatic diseases. The choice of treatment dictates a full knowledge regarding each disease and a through differential diagnosis. The aim of this manuscript is to review the subaxial cervical spine deformities.

Key Words: *Cervical spine, deformity.*

INTRODUCTION

Subaxial deformities of the cervical spine affects both children and adults. It can be due to degenerative, inflammatory, traumatic, or neoplastic conditions in the child and adult (Table 1). Biomechanically cervical spine faces a variety of loadings. Subaxial deformities tend to be progressive and needs surgical correction. Some of these deformites (e.g., deformity secondary to degenerative disc disease) develops slowly, whereas others (e.g., trauma) develops quickly. In most conditions, a correction and stabilization should be combined with a decompression. An understanding of cervical subaxial deformities dictates understanding of cervical spine biomechanics.

Table 1. The causes of subaxial deformities

Trauma
Degenerative disease
Tumors
Infections
Congenital deformities
Iatrogenic deformities
Post–laminectomy deformities
Post–irradiation deformities
Inflamatory disease
Ankylosing spondylitis
Rheumatoid arthritis
Neurofibromatosis
Rare causes of subaxial deformity

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Biomechanical aspects of cervical spine:

The initial curve of the spine is kyphotic. This kyphotic curvature remains in thoracic and sacral spine. To maintain sagittal balance, the normal cervical and lumbar spines become lordotic. The sagittal vertical axis passes through the middle of C1 and T1. Because of lordotic curvature, this axis passes dorsal to cervical spine vertebral bodies. A normal cervical spine sagittal alignment was reported –14.4° in the subaxial cervical spine (3). A decrease in cervical lordosis means straighthening or hypolordosis, and a revearse cervical curvature means kyphosis. All cervical osseo–ligamentous structures and musculature play important role in the cervical spine biomechanics. Cervical spine intervertebral discs and facet joints have determinant role for cervical spine stability. The ventral structures resist compressive forces, whereas the dorsal structures resist tensile forces (1, 18). Damage of either can change cervical spine sagittal balance, and may lead to subaxial deformities.

Conditions resulting in subaxial deformity:
Trauma

A variety of subaxial cervical trauma may cause deformity. These can be classified into 4 main groups, including; 1) flexion–dislocation; 2) flexion–compression; 3) compression–burst, and 4) extension injuries (1, 5, 22).

The most unstable form of subaxial traumas are flexion dislocations. They make up over 50% of all cervical injuries. A pure flexion injury leads to bilateral locked facet syndrome, whereas a combination of flexion and rotation leads to unilateral locked facet syndrome. This condition is associated with an intervertebral disc herniation as well as unilateral or bilateral facet capsule disruption. According to Nieminen, there is instability in almost 50% of patients with bilateral facet subluxation (5).

Flexion compression injuries lead to loss of less than 50–60% of the height of the vertebral body. The presence of vertebral body height loss greater than 30%, as well as the presence of dorsal ligamentous injury may be associated with instability and needs to be stabilized.

Compression burst fractures are secondary to axial loadings and are often unstable. The intrusion of bony fragments into the spinal canal dictates decompression and stabilization.

Extension injuries are secondary to dorsal directed forces and may cause dorsal rotation and/or translocation of the cervical vertebrae. In the hyperextension dislocation or sprain anterior longitudinal ligament and ventral disc disruption is common. The compression of dorsal elements may lead to laminar fractures. The management of simple hyperextension dislocations without neurological deficit is usually symptomatic. A severe form of hyperextension injuries, a hyperextension fracture–dislocation is commonly associated with injury of all dorsal elements, including articular masses, pedicles, laminae, and spinous process and is very unstable.

Posttraumatic subaxial deformities may be rarely seen many days after trauma despite the presence of normal radiographs at early posttraumatic stage (31).

Degenerative disease

Degenerative disease of the subaxial spine (e.g., cervical spondylotic myelopathy) may cause degeneration of both ventral and dorsal elements of subaxial cervical spine. Degeneration of subaxial cervical spine begins at ventral aspect of the intervertebral disc. It is characterized with loss of height of the ventral aspect of intervertebral disc and leads to loss of normal cervical lordosis. The straighthening of the cervical spine causes the increase in moment arm of the forces applied axially to the spine and, in turn, will cause the progression in kyphotic process. The presence of kyphotic cervical curve dictates a ventral approach for correction of cervical spine. The presence of straighthened spine may dictate the use of ventral or dorsal approach for stabilisation (1, 18, 22).

Tumors and infections

Spinal tumors and infections may also cause subaxial deformities. Vertebral metastases occur in about 5% of patients during the course of cancer. The majority of metastases (95%) are extradural, while 4.5% are intradural and 0.5% are intramedulary. Metastatic involvement of cervical spine predominantly affects the vertebral body. Spinal metastases most commonly causes lytic lesions in bone. Sclerotic bony involvement may also occur. Spinal instability was reported in 5 to 14% of patients with cervical metastases (16, 20). The main subaxial deformities include pedicle erosion, compression fracture, and pathologic fracture dislocations (10, 19). The involvement of intervertebral disc in metastases is not common, whereas it can be seen in infectious disease of the spine. The choice of treatment in most cases is surgery. Radiotherapy is usefull for patients with symptoms not due to instability. Spinal stability can be achieved with a variety of methods and materials. A patient with a good and prolonged survival needs to be stabilized with a bone graft and instrumentation, whereas a patient with short survival can be stabilized with methylmethacrylate and instrumentation.

Congenital deformities

Congenital deformities of cervical spine, which are caused by congenitally anomalous vertebral development are rare. They are seen in the growing child and adolescent and are divided into three main groups: defects of formation (such as hemivertebra); defects of segmentation (e.g., unilateral unsegmented bar); and combination of both (e.g., hemivertebra associated with a unilateral bar). Their cause usually is unkown. There are three major pattern: lordosis, kyphosis, and scoliosis (12, 25, 26, 30).

Embriologically, spinal column formation is observed at the 6th to 8th weeks of gestation. The centers of chondrification in the membraneous portions of mesenchyme begins to ossify and most of formation occur at this time. Wynne–Davies in a comprehensive review of the families of 377 patients, reported that an isolated anomaly such as hemivertebra carried no risk of a similar lesion in siblings or subsequent generation, whereas multiple level, complex anomalies carried a 5 to 10% risk of occurance (32).

There is few well-known environmental or biologic causes for congenital cervical deformities. The presence of spinal anomalies is often associated with the presence of tracheal, esophageal, renal, and auditory defects.

Most of these cases are young. Neck deformities occur commonly because relatively little normal spine above the area of segmentation defects, that is, little ability to develop a compensatory curve to balance the scoliotic portion. About 50% of cervical scoliosis are associated with Klippel–Feil anomalies. When scoliosis is associated with Klippel–Feil abnormalities, torticollis, or progressive lateral deviation of neck can be severe (30).

The pattern of unilateral unsegmented bar and contralateral hemivertebra with open disk space may be associated with poor prognosis, whereas the pattern of anterior block vertebra has the least likely prognosis for progression.

Iatrogenic subaxial deformities

Iatrogenic subaxial deformities include post–laminectomy and post–irradiation deformities. They are commonly seen in pediatric population in which their spine growth process in incomplete. However, an aggressive resection of dorsal osseous elements may cause an instability in adult patients as well (4, 8, 27, 28, 33, 35).

A. Post–laminectomy subaxial deformities.

Extensive laminectomy of cervical spine may lead to laminectomy–induced subaxial deformities. However, this condition is more commonly seen in pediatric population. A variety of reasons have been noted in the etiology of pediatric laminectomy–induced deformities. These reasons include ligamentous–osseous deficiency, neuromuscular imbalance, progressive osseous deformity (33). The importance of ventral and dorsal cervical columns in stability of cervical spine is known. In vertical compression, the ventral column carries 36% of the applied load, while each pair of facet joints transmit 32% of the applied load (1, 18). Dorsal osseoligamentous complex are stabilizer in tension, while ventral column is stabilize in compression. Extensive laminectomy destroy some of the dorsal structures such as facet joints, laminae, and dorsal musculo–ligamentous complex. As this deformity develops and progresses, the dorsal column is placed in tension and the ventral elements are placed in compression.

The true incidence of laminectomy–induced subaxial deformities is not known. Haft et al., reported a 59% incidence of kyphoscoliosis in children undergoing cervical laminectomy for spinal tumor (11). In an experimental study Lee and Moon compared the effect of multilevel laminectomy in immature and mature cats. They detected cervical kyphosis 4 months after laminectomy in immature cats group, whereas normal cervical curve was maintained in the mature cats group. They reported two main causes of postlaminectomy kyphosis; 1) subluxation of vertebral bodies; and 2) ventral wedging of the vertebrae (15).

The rate of laminectomy–induced subaxial deformity in adult population is lower than pediatric population, so that in the presence of normal cervical curvature and preservation of facet capsules, it is not commonly seen. The importance of preservation of facet joints during laminectomy was reported by many authors. Epstein reported the importance of facet joint contribution to stability and has recommended that not more than one quarter to one third of the facets be removed (7). Raynor et al., have shown that bilateral facetectomy of greater than 50% significantly reduces resistence to shear forces (21).

Zdeblick and Bohlman also reported a facetectomy greater than 50% causes a statistically significant instability in flexion and torsion (35).

Miyuzaki and co., reported a 17% incidence of deformity in adult following laminectomy for cervical spondylotic myelopathy (17). Mikawa reported a 11% rate of kyphosis following cervival laminectomy in the treatment of spondylosis (16). According to many authors the degree of facet resection correlates with subsequent deformity.

Goel et al., tested biomechanical effects of laminectomy and revealed 10% increase in range of motion, but did not feel that this was unstable (9). However, Saito et al., reported that following laminectomy tensile stresses distributed preoperatively in the dorsal ligaments were transfered to the facets in the postlaminectomy state. According to Saito et al., this led to an imbalance of stresses. This imbalance is severe in children because of relatively larger size of the head and its more ventrally located center of gravity (23).

Yasuka et al reported that postlaminectomy kyphosis can develop in children even if the facet capsules were preserved. According to them, two main causes of deformity in children is; hypermobility due to viscoelasticity of the ligaments, and wedging of the ventral vertebrae due to ventral compression of the cartilagenous endplates (33).

B. Post–irradiation subaxial deformities

Post–irradiation deformities are commonly seen in growing spine. Irradiation of the spine inhibites growth of the spine. Irradiation of whole spine leads to formation of smaller vertebral body than normal size. However, local irradiation of the vertebrae may cause different deformities. For example, irradiation of lateral part of vertebral body leads to growth inhibition of lateral portion of vertebral body and resultant deformity, whereas inhibition of ventral vertebral body by irradiation may lead to kyphotic deformity.

The main reason of deformity after irradiation of immature vertebrae is endplate involvement and its growth inhibition in axial plane. The degree of growth inhibition is related to the patient's age, and irradiation dosage. A does of 1000 rad does not produce a serious growth inhibition in endplate, whereas a permanent does of 2000 to 3000 rad or more may cause bone necrosis. The most severe bone changes following irradiation occur in children younger than 2 years. The initial post–irradiation changes are seen 6 months to 2 years after irradiation exposure. However, they manifest after many years (27).

Patients with mild deformities can be treated by orthosis. However, severe deformities needs to be stabilized using ventral or dorsal approaches. Pseudoarthrosis is common. Halo may reduce the risk of pseudoarthrosis.

Ankylosing spondylitis

Ankylpsing spondylitis (AS) is an inflamatory disease leading to periarticular fibrosis and ossification that may result in fixed kyphotic deformities of the spine. There are few cases with AS in whom kyphotic deformity occurs primarily in the cervical spine. This deformity is characterized by restriction of the field of vision, and interference with skin care under the chin and shaving in males. The weight of the head and gravity may pull the patient further into kyphosis. A combination of thoracic and cervical kyphosis results in "chin–on–chest" type of deformity. An important aspect of this disease is the presence of weakness of the posterior paraspinal muscles secondary to primary muscle disease. Facet inflamation may be painful and patient may tend to stand in a more flexed posture.

Acute fractures are usually seen in late stage AS when spines are completely fused. Seventy five percent fractures occur in the cervical spine secondary to hyperextension. Neurologic injury is severe The clinical picture in AS patient with spinal fracture may be complicated by the development of spinal epidural hematoma. Delay in diagnosis may result in a pseudoarthrosis. Therefore, a patient with AS should be radiographically investigated after any minor trauma. However, flexion–extension radiographs are not advised.

A positive HLA–B–27 cell surface antigen may help diagnosis. Although the HLA–B–27 is positive in 90% of patients with clinical AS, less than 2% of HLA–B–27 positive individuals will ultimately develop AS. Therefore this marker is useless in asymptomatic cases (27).

Simmons has postulated a C7–based extensive osteotomy of the cervical spine. Many authors advocated operation under local anesthesia (24). The use of general anesthesia is not common (13).

Rheumatoid arthritis:

Cervical spine involvement of rheumatod arthritis (RA) is not rare. It is observed in 36% to 88% of RA cases. Cervical spine is involved in three different patterns: (1): atlantoaxial subluxations; (2): cranial settling of odontoid; and (3) subaxial subluxations (SAS).

SASs are seen less common than the first 2 forms and may be combined with them. They are seen in 10% to 20% of RA cases. Subaxial abnormalities with RA include: multilevel SAS, subluxations below the previously fused segment, end plate erosions, ventral spondylitis, facet erosion, spinous process destruction, hyperlordotic deformities, epidural rheumatoid granulations, and axial shortening of the lower cervical spine. These signs may be missed in early stages. Subluxation are seen in C2–3 and C3–4 segments and have commonly "steplader" appearance (34).

Subaxial deformities commonly lead to spinal cord compression by pannus or by a pachimeningitis or arachnoiditis. It is a gradual process in which one vertebra slips with respect to another due to a combination of factors. Hughs reported the autopsy in two cases with SAS. Pressure on the ventral aspect of the spinal cord by a bony protrusion into the spinal canal produced cord lesions. The striking finding in each case was an irregular central area of partial infarction.

Lower cervical spine lesions in RA patients can produce intractable neck pain or myelopathy in association with destructive lesions in the cervical spine. Spine posture tends to show a gook–neck deformity in association with spinal canal stenosis and an increased range of motion of the lower cervical spine.

The treatment of choice in cases with neurologic deficit is surgery. A variety of ventral or dorsal decompression and fusions may be performed.

Neurofibromatosis:

Severe subaxial cervical deformities are quite rare in neurofibromatosis (NF). NF is an autosomal dominant genetic disorder that affects connective tissue derived from endoderm, ectoderm, and mesoderm. NF is classified as NF1 and NF2. Spinal deformities are the most common skeletal manifestation of NF–1. Majority of spinal deformities are located in the thoracic spine. Cervical spine involvement is unusual (29).

Dystrophic changes and osteolysis are often present at diagnosis. Cervical curve in NF1 is commonly kyphotic or kyphoscoliotic. The ventral wedging, scaloping, and displacement of the ventral body contributing to the acute angular kyphosis.

The other aspect of NF is the presence of intraspinal tumor. Tumor removal through laminectomy may lead to kyphotic deformity or accelerate progression of the preexistent dystrophic kyphosis.

There are few reports of cervical dystrophic kyphosis due to NF in the literature. Neck pain is the most common presenting complaint in NF–1 deformity of the cervical spine, but neurological complications including myeloradiculopathy, may occur.

The treatment of choice is surgery (14). Halo traction can be used for correction and postoperative immobilization in combination with two–stage ventral and dorsal fusion.

Rare causes of subaxial deformity:

Cuffe et al., reported 10 cases with dialysis–associated spondylarthropathy. Nine of them had a cervical instability and underwent a stabilization procedure. According to them, it was a result of hyperproduction of parathyroid hormone secondary to the abnormal metabolism of calcium and phosphate due to renal failure (6).

Bhorjraj et al., described a rigid cervical kyphosis in a case of adult–onset Still's disease, a chronic polyarthritis in children (2).

Treatment:

Because of the loads applied to the cervical spine, subaxial deformities tend to be progressive. Therefore, surgery is commonly indicated for; 1) decompression of spinal cord; 2) reduction; and 3) stabilization.

The operative decision–making process is affected by a variety of factors, including; 1) the presence of a fixed or reducible deformity, 2) The presence of spinal cord compression, and 3) the location of deformity (ventral versus dorsal, or three columns).

A fixed deformity with spinal cord compression requires a decompression and stabilization, whereas a fixed deformity without spinal cord compression rarely requires surgery for neck pain. The presence of reducible deformity requires reduction and a ventral fixation. A dorsal approach (e.g., lateral mass plating) is rarely indicated in subaxial deformities.

REFERENCES

1. Benzel EC: Biomechanics of spine stabilization. Principles and clinical practice. McGraw–Hill, Inc, New York, 1995.
2. Bhojraj SY, Dasgupta D, Dewoolkar LV: One–stage "front" and "back" correction for rigid cervical kyphosis. A new safer technique of correction for a rare case of Adult–onset Still's disease. Spine 18: 1904–1908, 1993.
3. Bridwell KH: Sagittal spinal balance–Symposium no. 1. Presented at AAOS section IV Scoliosis Research Society, New Orleans, Luisiana, February 1994.
4. Butler JC, Whitecloud III TS: Postlaminectomy kyphosis. Causes and surgical management. Orthop Clin North Am 23: 505–511, 1992.
5. Cooper PR: Stabilization of fractures and subluxations of the lower cervical spine. In Management of posttraumatic spinal instability. Neurosurgical topics. Edited by Cooper PR. AANS. Park Ridges, III, 1990, pp. 111–134.
6. Cuffe MJ, Hadley MN, Herera GA, Morawetz RB: Dialysis–associated spondylarthropathy. J Neurosurg 80: 694–700, 1994.
7. Epstein JA: The surgical management of cervical spinal stenosis, spondylosis, and myeloradiculopathy by means of the posterior approach. Spine 13: 864–869, 1988.
8. Farcy JPC, Weidenbaum M, Sola C: Surgical management of severe cervical kyphosis following extensive laminectomy. Spine 15: 41–45, 1990.
9. Goel VK, Clarck CR, Harris KG, Kim YE, Schulte KR: Evaluation of effectiveness of a facet wiring technique: An in–vitro biomechanical investigation. Ann Biomed Eng 17: 115–126, 1989.
10. Grote W, Kalff R, Kocks W, Schmit–Neuerberg KP, Pospiech J, Schax M: Operative treatment of metastatic cervical spine instability. In Advances in Neurosurgery Vol. 18, Edited by Bushe KA, Brock M, Klinger M: Springer Verlag, Berlin, 1990, 76–81.
11. Haft H, Ransohoff U, Carter S: Spinal cord tumors in children. Pediatrics 23: 1152–1159, 1973.
12. Hensinger RN: Congenital anomalies of the cervical spine. 16: 16–38, 1991.
13. Hyman SA, Rogers WD, Bullington III JC: Cervical osteotomy and manipulation in ankylosing spondylitis: Successful general anesthesia after failed local anesthesia with sedation. J Spinal Disord 3: 423–426, 1990.
14. Kokubun S, Ozawa H, Sakurai M, Ishii Y: One–stage anterior and posterior correction of severe kyphosis of the cervical spine in neurofibromatosis. A case report. Spine 18: 2332–2335, 1993.
15. Lee K, Moon M: The effect of multilevel laminectomy on the cervical spine of growing cats. An experimental study. Spine 18: 359–363, 1993.
16. Mikawa Y, Skihata J, Yamamuro T: Spinal deformity and instability after multilevel cervical laminectomy. Spine 12: 6–11, 1987.
17. Miyuzaki K, Tada K, Matsuda Y, Okuno M, Yasuda T, Murakami H: Posterior extensive simultaneous multisegment decompression with posterolateral fusion for cervical instability and kyphosis and/or S–shaped deformities. Spine 14: 1160–1168, 1989.
18. Naderi S, Benzel EC, Baldwin N: Cervical spondylotic myelopathy. Surgical decision making. Neurosurg Focus 1(6): article 1, 1996.
19. Perrin RG, McBroom RJ, Perin RG: Metasatic tumors of the cervical spine. Clin Neurosurg 37: 740–755, 1989.
20. Rao S, Badani K, Schildhaeur T, Borges M: Metastatic malignancy of the cervical spine. A nonoperative history. Spine 17 (10 Suppl): S. 407–412, 1992.
21. Raynor RB, Pugh J, Shapiro I: Cervical facetectomy and its effect on spine strength. J Neurosurg 45: 278–283, 1985.
22. Roy–Camille R, Saillant G, Laville C, Benazet JP: Treatment of lower cervical spinal injuries- C3 to C7. Spine 17 (10S): S. 442–446, 1992.
23. Saito T, Yamamuro T, Shika J, Oka M, Tsutsumi S: Analysis and prevention of spinal column deformity following cervical laminectomy I. Pathogenetic analysis of postlaminectomy deformities. Spine 16: 494–502, 1991.
24. Simmons EH, Bradely DD: Neuro–myopathic flexion deformity of the cervical spine. Spine 13: 756–762, 1998.
25. Smith MD: Congenital scoliosis of the cervical or cervicothoracic spine. Orthop Clin North Am 25: 301–310, 1994.
26. Smith MD, Lonstein JE, Winter RB: Congenital cervicothoracic scoliosis. A long term follow–up study. Orthop Trans 16: 165–170, 1992.
27. Spivak JM Giordano CP: Cervical kyphosis. In The Textbook of spinal surgery. Edited by Bridwell KH,

DeWald RL, Lippincott–Raven Pub, Philadelphia, 1997, pp 1027–1038.

28. Toyama Y, Matsumoto M, Chiba K, Asazuma T, Suzukii N, Fujimura Y, Hirabayashi K: Realignment of postoperative cervical kyphosis in children by vertebral remodeling. Spine 19: 2565–2570, 1994.

29. Ward BA, Harkey HL, Parent AD, McGuire RA, Kendig RJ: Severe cervical kyphotic deformities in patients with plexiform neurofibromas: Case report. Neurosurgery 35: 960–964, 1994.

30. Winter RB, Lonstein JE, Boachie–Adjei O: Congenital spinal deformity: Instruct Cours Lectures 45: 117–127, 1996.

31. Wilberger JE, Maroon JC: Occult posttraumatic cervical ligamentous instability. J Spinal Disord 3: 156–161, 1990.

32. Wynne–Davies R: Congenital vertebral anomalies: aetiology and relationship to spina bifida cystica. J Med Genetics 12: 280–288, 1975.

33. Yasuka S, Peterson HA, Laws ER, McCarthy CS: Pathogenesis and prophylaxis of postlaminectomy deformity of the spine after multiple level laminectomy: differences between children and adults. Neurosurg 9: 145–153, 1981.

34. Yonezawa T, Tsuji H, Matsui H, Hirano N: Subaxial lesions in rheumatoid arthritis. Radiographic factors suggestive of lower cervical myelopathy. Spine 20: 208–215, 1995.

35. Zdeblick TA, Bohlman HH: Myelopathy, cervical kyphosis and treatment by anterior corpectomy and strut grafting. J Bone Joint Surg (Am) 71: 170–176, 189.