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[REVIEW ARTICLE]

An Atypical Case Of Quadriparesis following C1 C2 Subluxation with Hydrocephalus: Effect of Extensive Rehabilitation towards Functional Independence. A Case Study.

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ABSTRACT :

Background: Atlantoaxial subluxation occurs when there is misalignment of the atlantoaxial joint. Several etiologies confer increased risk of Atlantoaxial subluxation in children, including neck trauma, inflammation, infection, or inherent ligamentous laxity of the cervical spine.

Methods: A case study was performed. Demographics and clinical data were reviewed for etiology, imaging techniques, treatment, and clinic outcomes.

Results: Patient suffered cervical spine trauma, advised for cervical collar after the surgery. In comparison to the baseline, the physiotherapists noted improvement in functional activities. Assessment and management of 1 patient with prolonged recovery from acquired hydrocephalus with VP shunting associated with C1-C2 subluxation.

Conclusion: Pediatric patient with atlantoaxial subluxation was a small case study with a single participant. Conservative treatment with hard cervical collar and immobilization after reduction may be necessary. Physiotherapy treatment benefitted for all the functional activities in her daily living.

Keywords: C1-C2 Subluxation, hydrocephalus & Physiotherapy treatment.

Introduction:

Hydrocephalus is a form in which excess cerebrospinal fluid (CSF) builds up within the fluid-containing cavities or ventricles of the brain. The term hydrocephalus is derived from the Greek words "hydro" meaning water and "cephalus" meaning the head. Although it translates as "water on the brain," the word actually refers to the buildup of cerebrospinal fluid, a clear organic liquid that surrounds the brain and spinal cord.

Hydrocephalus cases were regularly described by Hippocrates, Galen, and early and medieval Arabian physicians, who believed that this disease was caused by an extracerebral accumulation of water. Operative measures used in ancient times are neither proven by brain findings today clearly reported in the literature. Evacuation of superficial intracranial fluid in hydrocephalic children was first described in detail in the tenth century by Abulkassim Al Zahrawi.[1]

Hydrocephalus is a condition of cerebrospinal fluid, leading to enlargement of the ventricular system within the brain, typically associated with increased intracranial pressure. The prevalence of infant hydrocephalus is roughly one case per 1000 births. One of its most common causative mechanisms is congenital aqueduct stenosis, a focal reduction in the cerebral aqueduct at the level of the colliculus.[2]

The medical literature concerning neuropathological changes caused by hydrocephalus is reviewed. In humans the ependyma suffers focal destruction, cerebral blood vessels are distorted and capillaries collapse, there is damage to axons and myelin in the periventricular white matter, and occasionally neurons suffer injury. The damage seems to result from mechanical distortion of the brain combined

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with impaired cerebral blood flow. If ventriculomegaly develops very early, foci of cortical dysgenesis may be result. The appeal and distribution of pathological changes are dependent on the age at which hydrocephalus develops, the rate and magnitude of ventricular enlargement, and the duration of hydrocephalus. Diversionary shunting of cerebrospinal fluid can only incompletely reverse the damage and the potential for reversal diminishes as the duration of hydrocephalus increases.[3]

The most effective method used to investigate malformations such as Chiari II malformation would be ultra-sonography and MRI are than CT scanning (Fieggen et al., 2014). This leads to the two types which is congenital (detected at birth) and acquired (injury, illness and environmental factors) hydrocephalus.Study conducted by Gabriela Ramires De Oliveira (2020) on a 2-months-old Brazilian girl presented to the physiotherapy department diagnosed with congenital hydrocephalus. The intervention was based on the CME rehabilitation method, and its main principles of provoking active movements and minimizing handling [4].

1. Case Presentation:

1.1. Case History :

An 11 year old girl who had normal birth history presented to the physiotherapy department diagnosed with acquired hydrocephalus had past history of fever and neck injury for 15 days which was diagnosed with Atlanto - Axial Dislocation and MRI findings revealed significant anterior listhesis with cranial displacement of dens of C2 crossing the forearm magnum and indenting the lower part of medulla oblongata – Atlantiaxial dislocation , STIR hyperintensity seen in upper cervical cord and lower medulla–compressive myelopathy / contusion / oedema. STIR hyperintensity seen inC1 and C2 vertebral bodies and adjacent soft tissue and ligaments – oedema. STIR hyperintensity seen in bilateral paravertebral and prevertebral soft tissue – myofascial and TB since 6 months and was undergoing with medications for the same. Where she developed with inability to speak. Also got operated for atlantoaxial dislocation (C1 – C2 repair) which was done by metallic fixation device in suboccipital region and MRI findings revealed Moderate hydrocephalus and periventricular CSF ooze seen. Shunt tube seen in situ starting from posterior horn of right lateral ventricle extending and abutting lateral wall of left ventricle.

1.2. Clinical Assessment :

The patient was unconscious also was not oriented to time, place and person and had inability to speak. Her Glassgow Coma Scale Score was 6 (E4V1M1). Superficial, Deep and Cortical sensations could not assess as the patient was unconscious but patient did respond to pain stimulation. Deep tendon reflexes on both side of upper limb were exaggerated and lower limb were diminished whereas Babinski sign was positive bilaterally. She had grade 1 spasticity in both the elbow flexors and wrist extensors according to modified Ashworth scale. Tone was hypotonia for bilateral lower limb. There was restricted neck movement. There was no any bed mobility carried by patient.

1.3. Intervention :

Patient vitals were monitored before and after the rehabilitation training. Rehabilitation training was carried out for each day two sessions, six days a week excluding all the government holidays. Patient was initially advised for cervical collar after the surgery. She was given proper positioning to prevent bed sores. Coma stimulation program was started in which visual and auditory stimulation was given for 5s, twice with a 3s break material was used family photographs, video recordings of family and friends and religious chants using earphones. Rehabilitation training was followed with Specific, Measurable, Attainable or Assignable, Realistic, Time-related protocol.

Week 1:

Vitals were monitored periodically throughout the rehabilitation training. Coma stimulation was provided for visual stimulation material used was family photograph and video recordings of family and friends which was given for 5s twice with a 3s break and auditory stimulation was given for 5s – 10s twice with a 3s break on each side using earphone for religious chants. Proper positioning to prevent pressure sores was given. Bed mobility was given to the patient which included supine to side lying on either sides. Passive range of motion exercises were given to bilateral upper limb and lower limb. After week period the patient Glasgow Coma Scale Score was 8 (E4V1M3).

Week 2:

Vitals were monitored periodically throughout the rehabilitation training. The previous protocol was

continued with addition to bed mobility and passive range of motion to the upper limb and lower limb with neck movements, patient was also taken in long sitting position. Joint compression for elbow and wrist joint was given to normalize the tone. Oro motor stimulation was started with speech therapy. Each exercise was performed 10 times within the duration of 45 mins twice a day regularly except government holidays.

Week 3:

During the third week of rehabilitation training the patient Glasgow Coma Scale Score was 13 (E4V5M6) i.e. conscious state. The tone was normalized for elbow flexors and wrist extensors therefore elbow stretching and joint compression for wrist joint was discontinued. Active assisted range of motion was started for both the upper and lower limb, pelvic bridging with 5 sec hold x 5 repetition (fig 1) and patient was also taken to the high sitting position with support (fig 2).

Fig 1: Pelvic Bridging

Fig 2: High Sitting Position

Week 4:

During the fourth week the patient's Glasgow Coma Scale was 15 (E4V5M6). Patient was able to move her bilateral upper and lower limb actively with full range of motion. Strengthening was given to bilateral upper and lower limb with 1/2 kg weight cuff x 10 repetition for 2 sets/ day. PNF D1 flexion and extension pattern was given to bilateral upper and lower limb to increase the strength of the patient for 5 repetitions. To improve the dynamic sitting balance sitting reach outs was given to the patient with foot supported and for dynamic standing balance reach outs was given in standing position with the help of walker (fig3). Sit to stand was given with the help of walker (fig 4). Patient was ambulated with the help of walker and advice to walk with a 3-point gait (fig5). All the above treatment was given for twice a day.

Fig 3: Reach-Outs in Standing Position

Fig 4: Sit to Stand with Walker

Fig 5: Ambulation with walker

Week 5:

Patient did achieve all the bed mobility, bed side sitting to standing with minimal assistance, able to do all the functional activities from upper limb and lower limb, strengthening training was carried out with practicing task oriented exercise (specific to everyday movements e.g. Sitting to Standing, Independent walking, Stair climbing, Obstacle walking and Multi tasked activities.) fig 6&7. Each exercise was performed 10 times within the duration of 45 mins twice a day regularly except government holidays.

Fig 6: Stair Climbing

Fig 7: Independent walking with hands support

Exercise benefits the body generally in SB peripheral circulation, bowel function, reduces obesity, reduces sedentary lifestyle and urinary (Roussos et al., 2001) which are all challenges SB patients are likely to experience. Most articles reviewed are more oriented on mobility in relation to physiotherapy management which is an indication of its importance as one grows older (Bid, 2011).[4]

Discussion:

The purpose of this case study was to provide a framework of evaluation and subsequent treatment techniques for the assessment and management of patients with prolonged recovery from acquired hydrocephalus with VP shunting associated with C1-C2 subluxation. Posthemorrhagic hydrocephalus with ventriculomegaly is a serious sequela of IVH. The blood may spread through the ventricular system and accumulate in the basilar cisterns at the posterior fossa. Particulate blood clots may impair CSF circulation through the fourth ventricle and the sylvian aqueduct, and block reabsorption sites at the arachnoid villi through the development of adhesions in the basal cisterns.[6]

Kennedy et al reported that the use of acetazolamide and furosemide in preterm infants with PHH is ineffective in decreasing the rate of shunt placement and is associated with increased neurological morbidity. Other treatment modalities include serial lumbar punctures, serial ventricular punctures, the placement of a subcutaneous ventricular catheter reservoir or shunt, the insertion of an external ventricular drainage, and the placement of a V-P or ventriculosubgaleal shunt. [7]

Kawabe and associates concluded study on dens-facet angle of the axis was steeper in children than in adults, and meniscus-like synovial folds were found in the C0/1 and C1/2 facet joints of the spines of children but not in those of adults (Kawabe et al., 1989; Mercer and Bogduk, 1993)[8]. In Lin et al.'s study (1995), in the supine position, they showed that a 5 kg traction force increased both the intervertebral foramina area and disc height. In the three cases presented, the main components of the treatment procedure are cervical axial traction force and mobilization of subluxated joints. The axial maneuver force may have the effect to make some separation of the cervical joints, which could relieve the symptoms. In these three cases muscle spasm was found.[9] According to Grieve (1988), radiological examination is preferable before manipulation to

exclude organic disease.[10] However, Maitland (2001) states that "It is a cardinal rule that movements must never be forcibly thrust through protective spasm" and for that reason, grade V or thrust techniques on these patients was prohibited. Other absolute contradictions on cervical spine manipulation such as bone disease, inflammatory arthritis, positive Lhermitte's sign etc. should be carefully considered before manipulation[11]. In adult patients the three cases all got improvement immediately after manual therapy and achieved complete recovery within two days. A cervical orthosis was not prescribed. Although cervical collar is usually recommended for AARS, (Subach et al., 1998; Rahimi et al., 2003)[12] The major cause of immobilization is the spasticity which occurs in about 70 % of patients with SCI. It is also associated with the muscle paralysis, reduced muscle tone and absent tendon reflexes below the level of injury.[13] Physiotherapy focuses to make the patient independent as far as possible. To achieve strengthening of the weaker muscles. The devices used for some of the patients are also customized, sometimes designed and manufactured by our centre. They are used by patients for standing upright, balancing, standing and walking. Besides mobilization, they limit/remove the risk of deformities, overstretching and contractures, as well as maintain the anatomical/ functional position of the extremity. Approximately 60% of the neck rotation is from the atlantoaxial (C1–C2) joint combined with the horizontal C1–C2 facet joint alignment. Repetitive strain injury predisposes the cervical vertebrae to biomechanical changes and contributes to the development of C1–C2 subluxation.[14] Studies also report the advantages of orthosis for primary management of SCI which involve the benefits associated with standing, walking for short distances, etc.[15]

Conclusion:

Pediatric patient with atlantoaxial subluxation was a small case study with a single participant. Atlantoaxial subluxation (AAS) refers to the imperfect alignment of the C1-C2 vertebrae that may result from cervical trauma or precipitate from infection and consequent inflammation to the cervical tissue. Reduction of pain and swelling, improvement in motion, improvement in gross and fine motor skills, and relaxation and healing of soft tissues is the main focus of physiotherapy. Conservative treatment with hard cervical collar and immobilization after reduction may be necessary. Physiotherapy treatment benefitted for all the functional activities in her daily living. Which included hand activities, sit to stand, balance in sitting and standing with both static and dynamic, walking and stair climbing.

References:

1. Aschoff A, Kremer P, Hashemi B, Kunze S. The scientific history of hydrocephalus and its treatment. Neurosurgical review. 1999 Oct;22(2):67-93.
2. Tully HM, Dobyns WB. Infantile hydrocephalus: a review of epidemiology, classification and causes. European journal of medical genetics. 2014 Aug 1;57(8):359-68.
3. Del Bigio MR. Neuropathological changes caused by hydrocephalus. Acta neuropathologica. 1993 May;85(6):573-85.
4. Banda F. Physiotherapy management of spina bifida in Lusaka, Zambia.
5. de Oliveira GR, Fabris Vidal M. A normal motor development in congenital hydrocephalus after Cuevas Medek Exercises as early intervention: a case report. Clinical Case Reports. 2020 Jul;8(7):1226-9.
6. Hansen AR, DiSalvo D, Kazam E, Allred EN, Leviton A. Sonographically detected subarachnoid hemorrhage: an independent predictor of neonatal posthemorrhagic hydrocephalus?. Clinical imaging. 2000 May 1;24(3):121-9.
7. Kennedy CR, Ayers S, Campbell MJ, Elbourne D, Hope P, Johnson A, International PHVD Drug Trial Group. Randomized, controlled trial of acetazolamide and furosemide in posthemorrhagic ventricular dilation in infancy: follow-up at 1 year. Pediatrics. 2001 Sep 1;108(3):597-607.
8. Kulkarni AV, Warf BC, Drake JM, Mallucci CL, Sgouros S, Constantini S. Surgery for hydrocephalus in sub-Saharan Africa versus developed nations: a risk-adjusted comparison of outcome. Child's Nervous System. 2010 Dec;26(12):1711-7.
9. Kawabe N, Hirotani H, Tanaka O. Pathomechanism of atlantoaxial rotatory fixation in children. Journal of pediatric orthopedics. 1989 Sep 1;9(5):569-74.

10. Lin CJ, Lin RM, Wu MH. Atlantoaxial rotatory instability secondary to odontoid hypoplasia as a cause of acute torticollis in children: report of one case. Zhonghua Minguo Xiao er ke yi xue hui za zhi [Journal]. Zhonghua Minguo Xiao er ke yi xue hui. 1995 Nov 1;36(6):448-51.
11. Grieve GP. Common vertebral joint problems. Elsevier Health Sciences; 1988.
12. Maitland GD. Vertebral manipulation. Elsevier Health Sciences; 1986 Mar 20.
13. Subach BR, McLaughlin MR, Albright AL, Pollack IF. Current management of pediatric atlantoaxial rotatory subluxation. Spine. 1998 Oct 15;23(20):2174-9.
14. Bastian HC. On the symptomatology of total transverse lesions of the spinal cord; with special reference to the condition of the various reflexes. Medico-chirurgical transactions. 1890;73:151.
15. Ghanem I, El Hage S, Rachkidi R, Kharrat K, Dagher F, Kreichati G. Pediatric cervical spine instability. Journal of children's orthopaedics. 2008 Mar;2(2):71-84.
16. Campbell JH. Linked hip-knee-ankle-foot orthoses designed for reciprocal gait. JPO: Journal of Prosthetics and Orthotics. 2006 Jun 1;18(7):P204-8.