

Arnold-Chiari Malformation

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Definition

The usual age of onset of Arnold-Chiari malformation ranged from the second to the fourth decade, with female preponderance. It is assumed to rarely present clinically before adolescence¹ and is often associated with syringomyelia². Arnold-Chiari Malformation can be divided in three subtypes;

Type 1: (MC) Caudal herniation of the cerebellar tonsils through the foramen magnum that exceeds 5 mm into the cervical spinal canal.^{1,3}

Type 2: Caudal herniation that involves the cerebellar vermis and medulla, usually diagnosed in childhood with an average survival time of less than 2-3 years.⁴

Type 3: Caudal herniation of cerebellum and medulla accompanied by meningoencephalocele of the top of cervical vertebra and occipital region, cerebellar prolapse and hydrocephalus. ACM type III is an extremely rare anomaly with poor prognosis for newborn infants because of respiratory failure, swallowing dysfunction, hypertonía, or amyotonia.¹

Symptoms

The symptoms usually consist of neck ache, headache, weakness and numbness that present and progress gradually, but can be asymptomatic.^{1,3,5} Compression of the medulla, spinal cord and cerebellum or blockage of CSF can cause various symptoms: lower cranial nerve palsies, vertigo, visual and hearing disturbances, nystagmus, swallowing difficulties, spastic or ataxic paraparesis, bladder dysfunction, gait abnormality, tremor and chronic headache because of hydrocephalus.^{1,2}

DDX: Multiple sclerosis, primary headache syndromes, spinal tumours and benign intracranial HT.

Diagnosis

Diagnosis of the disease without radiologic evaluations is difficult because there are no distinguishing symptoms of the disease.⁴ Radiography may offer early suspicion for ACM: Small posterior fossa; Enlarged foramen magnum with a convex posterior border; Enlarged diameter of the spinal canal. MRI is the procedure of choice.

Treatment

Arnold-Chiari Malformations may be prevented by precon-

ceptional folic acid (or methylfolate if positive for the MTHFR genetic mutation) and Vitamin B12 supplementation.⁷ The ACM process usually begins during the third week of embryonic life at the time of closure of the neural groove.⁶

ACM with pregnancy

Women with ACM were not more likely to die during delivery-related admissions despite more frequent severe morbidity. They are more likely to develop severe medical complications including acute respiratory distress syndrome, stroke/cardiovascular accident, sepsis and seizures. Women with ACM were also more likely to be delivered by caesarean or develop preeclampsia.⁷

Medical treatment

The typical management of ACM is suboccipito-cervical decompression surgery. Surgery has a typical prognosis of greater than 80% improvement in ACM.⁴

Chiropractic literature

Few studies reported on chiropractic treatment for ACM patient. The first showed no improvement of symptoms for two patients with ACM.⁶ In the second, Applied kinesiology chiropractic and SOT treatment was used for treatment of loss of vision and nystagmus. After treatment, the patient's ability to see, read, and performs smooth eye tracking showed improvement.⁸ According to another study, asymptomatic Type I ACM is not necessarily a contraindication to skilled adjustments to the cervical spine.⁹ The frequency of complications after spinal manipulation is not known, but seem to be relatively risk free. One case study, showed significantly increased of symptoms after spinal manipulation of ACM patient. The literature would suggest that there is a real, if small, complication rate with a substantial long-term morbidity and disability.⁵

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Bell's palsy in infants, children and adolescents

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Bell's palsy defined as unilateral facial weakness is a relatively common pediatric problem affecting children from infancy to adolescence.^{1,2} Considered idiopathic,^{2,3} it is the most common cause of unilateral facial weakness.^{1,3} It is believed to be a post infectious, allergic, or immune neuritis affecting the facial nerve.¹ Bell's palsy is a common neurologic manifestation of Lyme's disease.¹ Other causes of facial nerve palsy include congenital, traumatic and neoplastic.¹ Clinical manifestation may have sudden onset within a period of hours and are: weakness of upper and lower face¹ (forehead muscle sparing is suggestive of UMN lesion due to bilateral innervation); inability to close eye; excessive tearing or excessively dry eyes; unilateral drooping of the corner of the mouth;² loss of nasolabial fold; normal sensation; lost of taste anterior two-thirds of tongue; difficulty in feeding because of impairment of sucking.⁷ Initial presentation may be pain around the ear and surrounding areas and associated with hyperacusis or dysacusis.¹

Bell's palsy is a diagnosis of exclusion and a careful history and examination will usually lead to a correct diagnosis.¹ The details of pregnancy, labour and delivery, maternal medical history and family history should be carefully taken.⁷ In birth trauma, risk factors include long pregnancy, long and/or difficult labor^{5,6,7} especially forceps delivery.^{5,6,7} birth weight of more than 3500g⁵ (may be seen if mother has diabetes⁶) or cephalopelvic disproportion,⁵ use of epidural anesthesia,⁶ use of a medication to cause labor and stronger contractions⁶ and primaparity.⁵ With infant or newborn, observation and examination for ecchymosis, bruising of scalp, hemotympanum, facial swelling, severe head molding^{5,7} and for Erb's palsy^{7,9} is needed.

Dysmorphic features, other cranial nerve palsies, other coexisting anomalies and family history of facial nerve

palsy or of other congenital anomalies favor developmental cause.^{5,7} Developmental causes include those associated with syndromes (such as Moebius syndrome) and teratogens.⁵ If a syndrome is suspected, referral for genetic testing⁷ may be necessary.

Facial palsy may develop at any time during childhood.⁴ Other differential diagnosis include infections¹ (Otitis media,¹ Mastoiditis,¹ Temporal bone abscess¹), trauma¹(head trauma)⁴, iatrogenic surgical injury,^{1,4} tumors⁴ (nerve tumors, leukemic invasion of facial nerve, rhabdomyosarcomas¹), stroke and infarcts.¹ Electrophysiology tests of facial nerve function can be done and is useful to assist with future surgical planning.⁵ Careful audiologic evaluation⁵ may also be recommended. Other tests may include X-Rays,⁹ CT Scan^{2,5} MRI^{2,3,5,6} (for trauma, temporal bone fracture, to rule out tumor or stroke).⁵ Lab testing may be used for Lyme's disease.¹ Some infants may have difficulty in feeding and may need additional support in establishment of feeding.^{5,7} If the paralysis does not resolve, it may affect the child's future speech, expressions of emotion and mastication. Referral to a speech and language therapist⁴ may be needed. Ophthalmologic evaluations may also be indicated.^{5,7} Plastic surgery⁷ referral may be considered for persistent, severe cases. Future psychosocial problems^{4,5} may appear for the older child and adolescents because of facial asymmetry, leading to a loss of balanced appearance and function.³ Recovery rates in infant, children and adolescents are high (85-93%)^{1,2,3} with complete recovery from weeks to months after onset.¹

Medical treatments include reassurance, corticosteroids or antivirals (Acyclovir)² and eye care (use of drops or artificial tears).^{1,2} Surgery is usually delayed until later in life⁵ (but risk of iatrogenic injury is high).⁵ Other treatment may

include acupuncture³ and physical therapy.³ The reported case⁸ of chiropractic care of an infant with traumatic right-sided facial nerve palsy (Bell's palsy) and right brachial plexus neuropraxia suggest that vast majority of cases (excluding severe cases) can be managed conservatively with manual therapy⁸ but also recommend close monitoring of the patient's signs and symptoms in order to determine if continuing to manage conservatively is appropriate.⁸ In chiropractic care of Bell's palsy, evaluation of upper cervical spine and cranial bones (occiput, temporal) for evidence of vertebral subluxation complex (VSC) should be performed with chiropractic adjustments administered where indicated.^{9,10} The motor branches of the facial nerve exit the skull through the stylomastoid foramen which is immediately behind the styloid process.^{9,10} A common cause of Bell's palsy could be compression neuropathy.⁹ VSC particularly in upper cervical spine and cranial bones could cause a compression neuropathy of the facial nerve.^{9,10} This might help to explain why correction of VSC in patients with Bell's palsy could produce restoration of facial muscle activity.^{9,10}

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Selective Mutism

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Selective mutism (SM) is characterized by a persistent failure to speak in specific contexts where speech is typically expected, despite normal hearing and speaking in other contexts. Onset of SM typically occurs before a child is 5-years-old (between ages three and six). But, it is usually first noticed when the child enters school (diagnosis occurs between ages five and eight). This disorder and specific features are described in the *2000 Diagnostic and Statistical Manual of Mental Disorders* (DSM-IV-TR) (pp.125-127) as listed below:^{1,2,3}

- Consistent failure to speak in specific social situations in which there is an expectation for speaking (for example at school) despite speaking in other situations (at home or in familiar context).
- The disturbance interferes with educational or occupational achievement or with social communication.
- The duration of the disturbance is at least 1 month (not limited to the first month of school).

- The failure to speak is not due to a lack of knowledge of, or comfort with, the spoken language required in the social situation.
- The disturbance is not better accounted for by a Communication Disorder (like stuttering) and does not occur exclusively during the course of a pervasive developmental disorder, schizophrenia, or other psychotic disorder.

SM can be present with a variety of comorbidities such as enuresis, encopresis, obsessive-compulsive disorder, depression, language abnormalities, developmental delay, and Asperger's disorders. The majority of children with selective mutism will outgrow the disorder spontaneously for unknown reasons. However, residual social phobia and other anxiety disorders may persist.⁴

Traditional treatment consist in two primary domains: psychotherapeutic approaches and medication-based interventions. Most common non-medication approaches to treat

SM are psychodynamic therapy (in children it's called individual play therapy), behavioral therapy, and family therapy. Within medication-based options, selective serotonin reuptake inhibitors (SSRIs) have been shown to improve mutism and anxiety. Results tend to be optimized when both approaches are employed simultaneously.

Social Communication Anxiety Treatment (S-CAT) is an evidenced-based treatment and is implemented at the Selective Mutism Anxiety Research and Treatment Center. S-Cat consists in a complete program which incorporate anxiety lowering techniques, methods to build self-esteem, tools to help with social comfort and communication progression. Individualized treatment plan needs to be developed based on every child specific needs and particularity of his disorder. Parent and teacher educations, and environmental changes (at home and at school) are essential to help the child overcome SM.

Since SM is a very rare condition, no literature was found on chiropractic and SM. Neuro-Emotional Technique (NET) is a branch of chiropractic and is defined has "a methodology of finding and removing Neuro Emotional Complexes (NECs). A NEC is defined as a subjective maladaptation syndrome adopted by the organism in response to a real or perceived threat to any aspect of its survival. NET has been described as a treatment designed to address negative distressing stimuli by removing these patterns by accessing

the nervous system via stimulation of the spine."⁵ (Specific phobia is a quite common anxiety disorder and literature was found on the potential impact of NET on anxiety level associated with this disorder. "Compared with the non-intervention control group, statistical analysis indicates a significant advantage for the NET group in regard to state anxiety/subjective distress, reported fear, and avoidant behavior."⁶ Since specific phobias may have similar impact on activities of daily living (can disrupt lives, limit work efficiency, reduce self-esteem, and strain relationships) it would be interesting to explore NET with the child diagnosed with selective mutism.

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