Dental Treatment Considerations for the Patient with Hydranencephaly: A Case Report

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Overview

Background

Hydranencephaly is a congenital post-neurulation disorder affecting the central nervous system during the second trimester of pregnancy. It is characterized by destruction of the cerebral hemispheres resulting in an empty cranial cavity that is replaced with cerebrospinal fluid, while the cranial vault and meninges remain intact. It is usually fatal before birth or shortly after. Symptoms include hydrocephalus, epilepsy, respiratory failure, and poor psychomotor development, among others.[3,4]

Etiology

Most commonly accepted theory → ischemic stroke due to bilateral occlusion of the internal carotid arteries during the neurogenic phase (middle cerebral artery may also be occluded). This results in the absence of structures perfused by the ICA/MCA, while structures perfused by the posterior cerebral artery and the basilar artery remain preserved (i.e. cerebellum, brainstem, thalamus, basal ganglia, choroids plexus).[4]

Other theories:

- Intrauterine infection → necrotizing vasculitis → hydranencephaly
- Hypoxia - caused by severe maternal carbon monoxide exposure
- Syndromic/genetic - mutation of NDE1 gene
- Leukomalacia - extreme form of leukomalacia resulting in multiple cystic cavities that become coalesced in the cranial cavity
- Toxic exposure - maternal exposure to toxins, such as cocaine, smoking, and estrogens
- Twin pregnancy - death of a twin in-utero may lead to the development and accumulation of embolic material from the deceased twin

Epidemiology

Although the incidence is reported as 1 in 10,000 to 1 in 5,000, it is usually fatal before birth or shortly after, as very few children with this condition survive infancy. Most cases are therapeutically aborted[4].

Males and females are affected equally[4].

Diagnosis and Treatment

Diagnosis

Hydranencephaly is most commonly diagnosed during pregnancy via routine ultrasound or MRI (as early as the 13th - 26th week of pregnancy when the hemispheres and falx have been formed). If it is not diagnosed during pregnancy, diagnosis may be delayed from weeks to months as patients may initially present as normal. Within a few weeks, these patients develop hyperirritability, hyper/hypotonia, increase in head circumference, and wide open anterior fontanelles. Other signs and symptoms include: failure to thrive, spastic diplegia, cognitive delay, hypertelorism, vision and hearing problems.[3,4]

Differential Diagnosis

Severe-extreme hydrocephalus - buildup of fluid in the ventricles resulting in abnormal head circumference at birth
Alobar holoprosencephaly - incomplete division of the brain into right and left hemispheres resulting in facial anomalies and a small head circumference
Severe open schizencephaly - abnormal migration of neurons resulting in irregular gaps in the brain
Anencephaly - defects in the skull and brain[3]

Treatment

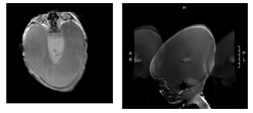
There is no cure for hydranencephaly. Treatment entails supportive management of symptoms. Prognosis is poor - most patients die in-utero. If patients make it to birth, most do not survive past the first year of life. Given the poor prognosis, if the diagnosis is confirmed during pregnancy, the option for a therapeutic abortion is often discussed.[4]

Management of Symptoms

- Hydrocephalus - ventriculoperitoneal or ventriculoatrial shunt to reduce intracranial pressure
- Seizures - anti-epileptic drugs
- Respiratory failure - tracheostomy and/or mechanical ventilation
- Poor psychomotor development - physical therapy
- Nutritional intervention[3]

Case Report

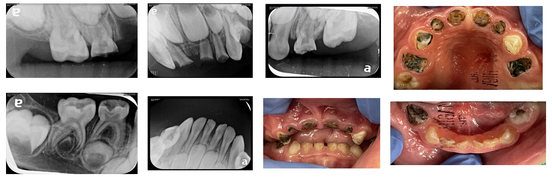
Patient Description

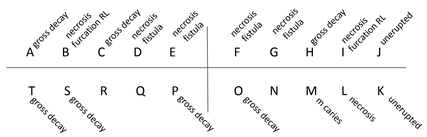


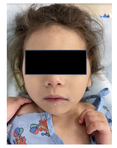
4y9m female Weight = 9.4kg
Health History: Hydrancephaly, hydrocephalus, optic atrophy, precocious puberty, partial diabetes insipidus, central adrenal insufficiency, chronic kidney disease, developmental delay
Medications: DDAVP, gabapentin, hydrocortisone, melatonin, simethicone, polyethylene glycol. Allergies: Latex, fentanyl
HPI: Patient presented to the ED due to fever, increased irritability, and several episodes of emesis. Sepsis was suspected due to leukopenia, bradycardia, and tachypnea. Hospital dentistry was consulted due to several dental abscesses being deemed as the source of infection.
Assessment: Severe early childhood caries; pulpal necrosis of maxillary central and lateral incisors; dental abscess of maxillary central incisors.
Plan: Given concern with fever and sepsis of unknown origin, urgent dental treatment was indicated under general anesthesia in the OR

Dental Treatment under GA

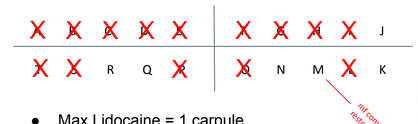
- Stress dose of steroids administered due to adrenal insufficiency
- Exam findings:

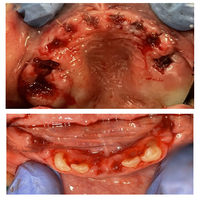






Treatment completed: 14 extractions, 1 composite restoration





- Max Lidocaine = 1 carpule
- Gelfoam in all sockets
- Single 3.0 interrupted chromic gut sutures in the areas of teeth #A, B, D, E, F, G, H, I, L, S, and T

Prognosis and Outcomes Assessment

Literature relating to dental treatment for patients with hydranencephaly is limited. Most patients do not survive long enough to require dental treatment, let alone for their teeth to erupt. After dental treatment in the OR, our patients remains with 4 primary teeth (#M, N, Q, and R; teeth #J and #K are still unerupted). Radiographs show that permanent teeth are present. Frequent recalls with dentistry will allow for close monitoring in order to prevent future dental infections.

Given the patient's medical condition and questionable prognosis, quality of life is the main focus. Our goal was to eliminate any dental source of infection that could potentially contribute to or cause sepsis, thus treatment was aggressive. Consequences of premature loss of primary teeth include difficulty chewing and ectopic eruption of permanent teeth.

Odontogenic infections may become severe if untreated, however, it is rare to have a bacteremic load large enough to lead to sepsis[2]. Literature search by Dave et al from January 1990 to December 2019 revealed 4 studies that identified sepsis from dental causes[1]. Although sepsis from a dental infection has been rarely reported, it is still a reality. ER physicians should consider the possibility of a dental infection causing sepsis when determining a potential cause for sepsis, even when it may not be manifested as an obvious extraoral swelling

References

1. Dave M, Barry S, Coulthard P, et al. An evaluation of sepsis in dentistry. Br Dent J. 2021;230:351-357.
2. Holmberg P, Hellmich T, Homme J. Pediatric Sepsis Secondary to an Occult Dental Abscess: A Case Report. J. Emerg. Med. 2017;52(5):744-748.
3. Pavone P, Pratico AD, Vitaliti G et al. Hydranencephaly: cerebral spinal fluid instead of cerebral mantles. Ital. J. Pediatr. 2014;40:79
4. Sandoval JI, De Jesus O. Hydranencephaly. [Updated 2022 Jun 27]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK558991/