Triple neural tube defect

The coexistence of three neural tube defects (NTDs) in a single child is an exceptional event. A review of the literature revealed nine published “double” NTD cases, but no cases of “triple” NTDs have been reported till 2005.

Case report

2016

A 1-year-old child with contiguous myelomeningocele and lipomyelomeningocele centered on Type I split cord malformation with Chiari II malformation and hydrocephalus. This composite anomaly is probably due to select abnormalities of the neurenteric canal during gastrulation, with a contiguous cascading impact on both dysjunction of the neural tube and closure of the neuropore, resulting in a small posterior fossa, probably bringing the unified theory of McLone closer to the unified theory of Pang [1].

2005

The rare case of a two-year-old boy with three distinct NTDs is presented. The boy had a 17x15x15-cm(3) parieto-occipital encephalocele, a small cervical myelomeningocele, and a 11x11x8-cm(3) thoracolumbar myelomeningocele. Hydrocephalus and Chiari II malformation accompanied the NTDs. All three lesions were surgically treated with good cosmetic results and satisfactory neurologic outcome.

Current neural tube closure theories and models are reviewed in an attempt to better understand this extremely unusual coexistence. The multi-site closure model is clearly more useful in our understanding of NTDs [2].

1)
Dhandapani S, Srinivasan A. Contiguous triple spinal dysraphism associated with Chiari malformation Type II and hydrocephalus: an embryological conundrum between the unified theory of Pang and the unified theory of McLone. J Neurosurg Pediatr. 2016 Jan;17(1):103-6. doi: 10.3171/2015.6.PEDS15179. Epub 2015 Oct 16. PubMed PMID: 26474100.
2)
Tekkök IH. Triple neural tube defect–cranium bifidum with rostral and caudal spina bifida–live evidence of multi-site closure of the neural tube in humans. Childs Nerv Syst. 2005 Apr;21(4):331-5. Epub 2004 Sep 29. PubMed PMID: 15455247.