

Open neural tube defect

Open [neural tube defects](#) occur when the [brain](#) and/or [spinal cord](#) are exposed at birth through a defect in the [skull](#) or [vertebra](#).

Examples

[Anencephaly](#)

[Encephalocele](#)

[Hydranencephaly](#)

[Iniencephaly](#)

[Schizencephaly](#)

[Spina bifida](#).

In most foetuses with [open neural tube defect](#), the end portion of cerebellum was below the [Occiput dens line](#) (ODL) associated with different degrees of [ventriculomegaly](#).

The proposed innovation aims to bring to the ultrasound the most likely anatomical parameters of evaluation in normal foetuses and in foetuses with [spinal dysraphism](#) ¹.

[Controversy](#) exists regarding the optimal mode of [delivery](#) for [fetuses](#) with open neural tube defects.

To compare neurological outcomes among infants with open neural tube defects who underwent vaginal compared to caesarean delivery.

Electronic databases [MEDLINE](#), [EMBASE](#), [Scopus](#), and [Clinicaltrials.gov](#) were searched from inception to November 2017.

Eligible studies included observational or randomised studies comparing vaginal and caesarean delivery in pregnancies with fetal open neural tube defects who did not undergo prenatal repair.

Two reviewers independently reviewed abstracts and full text articles. Outcomes were compared between vaginal and caesarean delivery and prelabour caesarean versus labour. The primary outcome was motor-anatomic level difference. Secondary outcomes included shunt requirement, sac disruption, meningitis, and ambulation at 2 years. Meta-analysis was performed and mean difference or odds ratios with 95% confidence interval calculated.

Of 201 abstracts identified in the primary search, 9 studies (672 women) met eligibility criteria. Comparing vaginal and caesarean delivery, there was no significant difference in motor-anatomic level difference (mean difference -0.10, 95% CI -0.58-0.38; I² =57%). The vaginal delivery group was less likely to require a shunt or have sac disruption (OR 0.37, 95% CI 0.14-0.95 and OR 0.46, 95% CI 0.23-0.90, respectively). Comparisons by prelabour caesarean versus labour showed no significant

difference in motor-anatomic level difference (OR 1.29, 95% CI -0.63-3.21) or ambulation at 2 years (OR 2.13, 95% CI 0.35-13.12).

Caesarean delivery was not associated with improved neurological outcomes among fetuses with open neural tube defects ²⁾.

Treatment

An open neural tube defect (ONTD) features an exposed, unclosed neural plate in the form of an expanded and frequently hefty neural placode. Traditional philosophy of ONTD repair aims at preserving the placode at any cost, which often means stuffing the entire thick and unwieldy but non-functional tissue into a tight dural sac, increasing the likelihood of future tethering of the spinal cord. The same philosophy of attempting to save the whole perimetry of the placode also sometimes leads to inadvertent inclusion of parts of the squamous epithelial membrane surrounding the placode into the reconstructed product, only to form inclusion dermoid cyst causing further injury to the neural tissues. Lastly, unsuccessful neurulation of the caudal primary neural tube almost always adversely affects junctional and secondary neurulation resulting in a defective conus, often with a locally active sacral micturition centre that is isolated from and therefore lacking suprasegmental inhibitory moderation. This frequently leads to the development of a spastic, hyperactive, low-compliance and high-pressure bladder predisposing to upstream kidney damage, without benefits of normal bladder function. We are introducing a new surgical technique designed to minimise or eliminate these three undesirable complications of conventional ONTD closure ³⁾.

Case series

In a retrospective cohort study of 47 patients with open neural tube defect (NTD) underwent prenatal repair (17 fetoscopic; 30 open-hysterotomy). At referral and six weeks postoperatively, the degree of [hindbrain herniation](#), ventricular atrial widths and ventricular volume were evaluated by MRI. [Head circumference](#) and ventricular atrial widths were measured on [ultrasound](#) at referral and during the last [ultrasound](#) before delivery. Anatomic level of the lesion (LL) was determined based on the upper bony spinal defect detected by ultrasound. Corroenne et al. considered the functional level as worse than anatomical level at birth when the motor level was equal or worse than the anatomical LL.

26% (12/47) of the cases showed worse functional level than anatomical level at birth. Having a HBH below C1 at the time of referral was associated with a worse functional level than anatomical level at birth (OR=9.7, CI95[2.2-42.8], $p<0.01$). None of the other brain parameters showed a significant association with motor outcomes at birth.

HBH below C1 before surgery was associated with a worse functional level than anatomical level at birth ⁴⁾.

¹⁾

de Sá Barreto EQ, Moron AF, Milani HJ, Hisaba WJ, Nardozza LM, Araujo Júnior E, Cavalheiro S. The occiput-dens line: the purpose of a new ultrasonographic landmark in the evaluation of the relationship between the foetal posterior fossa structures and foramen magnum. Childs Nerv Syst. 2015 May;31(5):729-33. doi: 10.1007/s00381-015-2621-x. Epub 2015 Feb 21. PubMed PMID: 25700613.

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3)

Eibach S, Pang D. A New Surgical Paradigm for Postnatal Repair of Open Neural Tube Defects Using Intraoperative Neurophysiology Monitoring. Adv Tech Stand Neurosurg. 2023;47:49-64. doi: 10.1007/978-3-031-34981-2_3. PMID: 37640872.

4)

Corroenne R, Zarutskie A, Guimaraes C, Yopez M, Torres P, Shetty A, Lee W, Espinoza J, Shamshirsaz A, Nassr AA, Belfort M, Whitehead, Magdalena Sanz Cortes. Is ventriculomegaly and hindbrain herniation seen before and after prenatal neural tube defect repair associated with a worse functional level than anatomical level at birth? Prenat Diagn. 2021 Jun 27. doi: 10.1002/pd.6000. Epub ahead of print. PMID: 34176146.

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