Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Gynaecology

Selective Twin Deformity in Di-Chorionic Twins: A Case Study

Ngawa Edith Ngalande[1], Fatima Zohra Fdili Alaoui[1], Sofia Jayi[1], Abraham Alexis Sanoh[1*], Hikmat Chaara[1], Moulay Abdelilah Melhouf[1]

[1]University Teaching Hospital Fez, Sidi Mohamed Ben Abdellah University of Fes, Morocco

DOI: https://doi.org/10.36347/sjmcr.2024.v12i12.047 | **Received:** 17.11.2024 | **Accepted:** 26.12.2024 | **Published:** 28.12.2024

***Corresponding author:** Abraham Alexis Sanoh
University Teaching Hospital Fez, Sidi Mohamed Ben Abdellah University of Fes, Morocco

Abstract **Case Report**

Neural tube defects (NTD) are a group of congenital malformations of the brain and spine, the etiology of which is still debated. Although presumed to be the consequence of interactions between genetic and environmental factors, so far, it is not known which genes are involved in the pathogenesis of these malformations. Dichorionic twin pregnancy discordant for fetal hydrocephalus (HC) with spina bifida is a serious condition that threatens the normal co-twin's life by causing foetal growth restriction, preterm labor and sudden death of one or both of the fetuses. This is a case report on twin pregnancy with fetal hydrocephalus and spina bifida associated with congenital hip dysplasia and a club foot, recognized in the 32th week of gestation. Course of pregnancy and management during labour are described. The peculiarity of our observation is the importance in establishing the significance of environmental factors as well as a genetic basis in NTD. Some occupational and other exposures, including maternal use of antiepileptic drugs (AEDs), are associated with increased risk for NTDs. Among women who have had an NTD-affected pregnancy, recurrence risk is markedly higher than the risk for a first NTD-affected pregnancy in the general population. There is strong evidence, overall, for a protective effect of adequate folate consumption. In some high-risk groups, however, such as women taking AEDs, folate supplementation has not been proven to reduce NTD risk.

Keywords: NTD, Hydrocephalus, Spina Bifida, Twin Pregnancy, Congenital Anomaly.

Copyright © 2024 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution **4.0 International License (CC BY-NC 4.0)** which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

Neural tube defects (NTDs) are malformations secondary to abnormal neural tube closure that occur between the third and fourth weeks of gestational age. They result in structural defects that occur anywhere along the neuroaxis from the developing brain to the sacrum and often result in the exposure of neural tissue [1].

HC is a dynamic disorder of cerebrospinal fluid production, flow and or resorption; it is however far more complicated than a simple disorder of CSF circulation. Although commonly considered a single disorder, HC is a collection of a heterogeneous complex and multifactorial disorders. It has an estimated incidence of 1 in 1500 births [2].

Number of specific chromosomal or single-gene disorders, presumably not affected by environmental influences, are associated with the development of NTDs, but such syndromal cases account for a small proportion of NTDs in live-born infants.

Congenital hydrocephalus is one of the commonest congenital anomalies of the central nervous system. It is characterized by extensive accumulation of cerebrospinal fluid within the ventricles of the brain due to an imbalance between synthesis and absorption of cerebrospinal fluid.

Dichorionic twin pregnancy discordant for fetal hydrocephalus (HC) with spina bifida is a serious condition that threatens the normal co-twin's life by causing foetal growth restriction, preterm labor and sudden death of one or both of the fetuses. This is a case report on twin pregnancy with fetal hydrocephalus and spina bifida associated with congenital hip dysplasia and a club foot, recognized in the 32th week of gestation. Course of pregnancy and management during labour are described below.

Patient History

A 33year old multiparous parturient presented to our maternity ward at 32 week gestation in the latent phase of labor, When the mother's anamnesis was questioned, it was learned that there was no toxin exposure during pregnancy, and she did not use drugs or

cigarettes. But patient did not attend prenatal checkups nor did she receive folic acid no prenatal vitamins during the course of her pregnancy, There was no history of diabetes no high blood pressure or pre-eclampsia. But no blood tests were performed to exclude either.

Clinical Findings and Diagnostic Approach

Upon vaginal examination it was found that the presentation was breach at 2cm dilated+ with an intact amniotic sac. Two sacs were seen in the Ultrasonography (USG) performed with twin 1 breach presenting with triventricular hydrocephalus and cranial perimeter of 37cm, Twin 2 cephalic presenting showing no abnormality at the limit of the ultrasound exam.

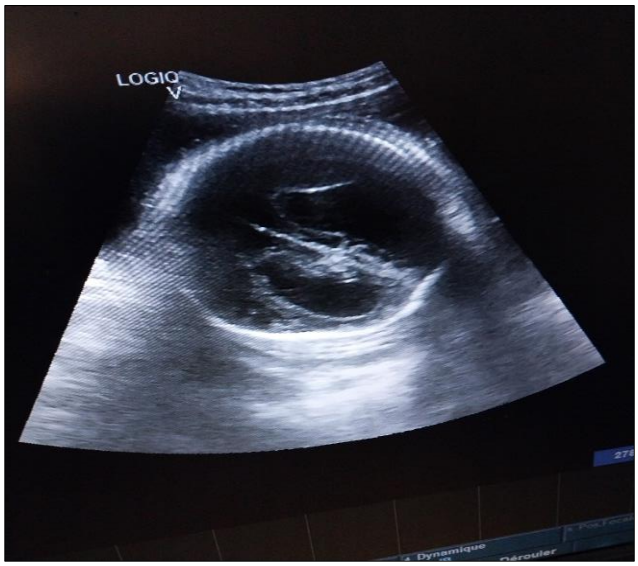

Figure 1: In utero foetal ultrasound showing Tri ventricular hydrocephalus

Therapeutic Intervention and Follow-Up

A caesarian section was performed and delivered two female babies. The Birth weights were 2400g (twin 1) and 1800g (twin 2), respectively which were both more than 2 standard deviations above the estimated values for the gestational age. While (twin 1) presented upon post birth examination hydrocephalus, a club foot with congenital hip displasia and spina bifida cystica with mengingocelle. (fig2,3,4). Her head circumference increased 1 cm in 3 days and the anterior fontanelle was bulging. Cranial ultrasonography revealed ventricular enlargement with no sign of hemorrhage. Computerized tomography was performed, and triventricular hydrocephalus was demonstrated. Soon after, the patient experienced a convulsion. Phenobarbital infusion was initiated and emergency surgery was performed to insert a ventricular reservoir. Intermittent CSF removal was performed for about 2 weeks, until her general status improved and she was free of fever. Afterwards she was operated on and a ventriculoperitoneal shunt was inserted while the ventricular reservoir was removed. She went on to be admitted in the in the pediatric surgery department for reparation of the spina bifida and hip dysplasia and club foot. (Twin 2) experienced an uneventful delivery. Her physical examination was within normal limits was followed up on an incubator for one month due to low birth weight and weak reflexes.

Figure 2: Foetus showing spina bifida cystica with meningocele

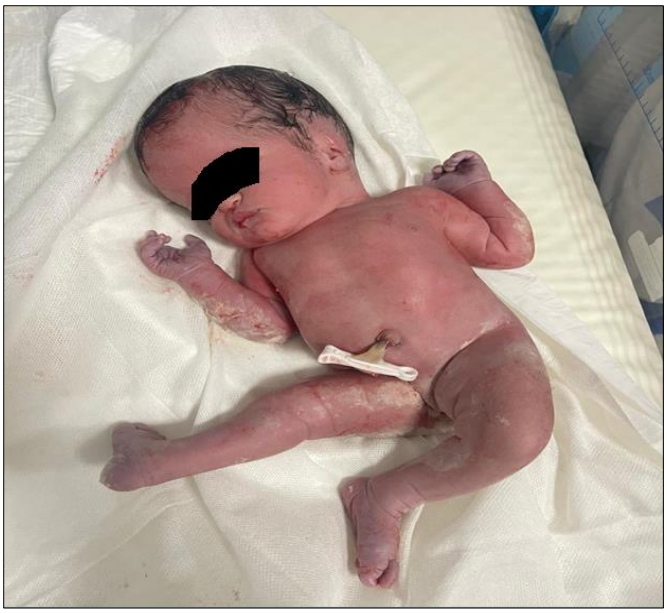

Figure 3: Foetus showing congenital hip dysplasia

Figure 4: Twin 1 presenting macrocephaly secondary to hydrocephalus in comparison with twin 2.

DISCUSSION

The concurrence of hydrocephalus and spinal bifida is not uncommon according to james kuhn *et al*., [3], as since anatomical relations of the brain and spinal chord allow for cerebral spinal fluid to communicate as is found in chiari II malformation. Recently, the advancement of prenatal screening and diagnostic tools for pregnant women ameliorates early detection of congenital anomalies. Literatures reported that hydrocephalus is the easiest congenital anomaly to be diagnosed using ultrasonographic examination [4].

In the NTD twin studies published to date, if the zygosity of the twin pairs is unknown, making exact conclusions about concordance difficult. Instead, twin pairs are usually divided into groups based on whether the twins are like-sexed or unlike-sexed, and these categories are used as surrogate markers for zygosity. In their synthesis of pooled data, Elwood *et al*., [5], found that the difference in concordance rates between like-sexed and unlike-sexed twins was larger for spina bifida than for anencephaly, but the difference was statistically significant only for all NTDs combined (like-sexed, 7.7 vs. unlike-sexed, 4.0) [1].

There are two forms of hydrocephalus, the congenital and the acquired. Congenital hydrocephalus is an aetiologically diverse disorder, including environmental (infectious, teratogenic,) and genetic factors as the underlying mechanism [2], underlying cause of acquired hydrocephalus (HC) is usually quite clear. On the other hand, the aetiology of congenital hydrocephalus is not well established. It is thought that congenital HC may develop at an important and specific embryonic time period of neural stem cell proliferation and differentiation in the brain [6]. Congenital HC may occur alone (nonsyndromic) or as part of a syndrome with other anomalies (syndromic) as was the case in our

patient who presented other deformities in addition to the NTD.

In syndromic forms, it may be hard to define the defective gene because of the association with other anomalies. Congenital HC may be associated with chromosomal abnormalities for example trisomy 13, trisomy 18, triploidy, Mendelian conditions such as Walker-Warburg syndrome, Meckel syndrome, Fanconi anemia, other central nervous system (CNS) malformations such as Dandy-Walker malformation, Chiari malformation, neural tube defects, or it may be isolated [7], as in our case where the foetus developed spina bifida and associated hip dysplasia.

The role of environmental factors in the pathogenesis of congenital hydrocephalus is also well documented in the literature [8]. Prenatal infections such as toxoplasmosis, rubella, syphilis and cytomegalovirus seem to demonstrate the strongest association [9]. The risk of hydrocephalus may be increased by the maternal consumption of drugs containing misoprostol, dextromethorphan, nalidixic acid, cephalosporins, for example [10, 11]. Maternal smoking does not appear to affect NTD rates. Although the association between hydrocephaly and maternal diabetes is not clear, a recent study demonstrated an increased risk among infants born to obese but non-diabetic women [1].

Once a mother has had a child with an NTD, the recurrence risks are markedly higher than reported population risks of a first NTD-affected pregnancy from the same countries during the same time period. The recurrence risk may increase after each affected pregnancy [1]. The effect of maternal age on risk of NTDs is generally considered to be small. When an association with age can be found, risk tends to be elevated in older or very young mothers. In a randomized trial of prevention of *recurrent* NTDs, folate reduced the risk by 75%, whereas multivitamins alone were associated with a 20% reduction in recurrence [12].

CONCLUSION

In a case of di-chorionique same sex twins such malformations are probably congenital. Concordance for NTD is likely if the twins are like sex and identical. Congenital hydrocephalus and other NTD seem to be a multifactorial disorder, triggered by environmental factors in genetically predisposed individuals. There are many clues to their etiology, and there is some evidence for a preventive intervention, folate supplementation, that may be effective for some individuals at risk.

Conflicts of Interest: The authors declare no conflicts of interest.

Author Contributions

All authors participated in the care of the patient, and in the writing and correction of the manuscript. They all also declare having read and approved the final version of the manuscript.

REFERENCES

1. Frey, L., & Hauser, W. A. (2003). Epidemiology of neural tube defects. *Epilepsia*, *44*, 4-13.
2. *East and Central African Journal of Surgery*, *14*(1), March-April 2009, 64-68
3. Kuhn, J., Weisbrod, L. J., & Emmady, P. D. (2024). Chiari Malformation Type 2. [Updated 2024 Feb 25]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; https://www.ncbi.nlm.nih.gov/books/NBK557498/
4. Abebe, M. S., Seyoum, G., Emamu, B., & Teshome, D. (2022). Congenital hydrocephalus and associated risk factors: an institution-based case–control study, Dessie town, North East Ethiopia. *Pediatric health, medicine and therapeutics*, 175-182.
5. Elwood, J. M., Little, J., & Elwood, J. H. (1992). *Epidemiology and control of neural tube defects*. Oxford University Press.
6. Adams, C., Johnston, W. P., & Nevin, N. C. (1982). Family study of congenital hydrocephalus. *Developmental Medicine & Child Neurology*, *24*(5), 493-498.
7. Schrander-Stumpel, C. T. R. M., & Fryns, J. P. (1998). Congenital hydrocephalus: nosology and guidelines for clinical approach and genetic counselling. *European journal of pediatrics*, *157*, 355-362.
8. Schrander-Stumpel, C. T. R. M., & Fryns, J. P. (1998). Congenital hydrocephalus: nosology and guidelines for clinical approach and genetic counselling. *European journal of pediatrics*, *157*, 355-362.
9. Stoll, C., Alembik, Y., Dott, B., & Roth, M. P. (1992). An epidemiologic study of environmental and genetic factors in congenital hydrocephalus. *European journal of epidemiology*, *8*, 797-803.
10. Czeizel, A. E., Rockenbauer, M., Sørensen, H. T., & Olsen, J. (2001). Use of cephalosporins during pregnancy and in the presence of congenital abnormalities: a population-based, case-control study. *American journal of obstetrics and gynecology*, *184*(6), 1289-1296.
11. Orioli, I. M., & Castilla, E. E. (2000). Epidemiological assessment of misoprostol teratogenicity. *BJOG: An International Journal of Obstetrics & Gynaecology*, *107*(4), 519-523.
12. Centers for Disease Control and Prevention (CDC. (1995). Prevention program for reducing risk for neural tube defects--South Carolina, 1992-1994. *MMWR. Morbidity and mortality weekly report*, *44*(8), 141-142.