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Research Article

Neural Tube Malformations: Monocentric Results of Operated Cases

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Abstract

Introduction: NTDs are malformations of the central nervous system. It's therapeutic complexities and the functional future of the child are the challenges in their management. The main objective of this study is to identify the epidemiological profile and short-term prognosis of these abnormalities.

Methods: This is a retrospective, descriptive study over a period of four years.

Results: The frequency of NTDs was of the order of 10 cases/year. The male gender was the most affected with a sex ratio of 2. The most common anatomoclinical form was Meningocele (53.65%). The association with congenital hydrocephalus found in this study was 26.82%. Nearly thirty-four percent of cases were children born of younger mothers, and the prenatal consultation rate was effective for 51.21%. Maternal infection was found in 58.53%. Folic acid intake during pregnancy was found in 82.73%. Surgery repair of the deformity was generally successful. Meningitis was the main complication. There were 4.87% of deaths related to serious cardiopulmonary diseases.

Conclusion: A clear male predominance of this condition, a significant history of maternal infection, the majority of anterior meningocele type topography, specify our series. Surgical outcomes are generally good except for a few manageable infected cases. The periconceptional and prenatal regular consultations are to be sensitized in order to prevent and reduce these abnormalities.

Keywords: Folic Acid; Prenatal Ultrasonography; Surgery; Tube Neural Defects

Introduction

The term "neural tube closure disorder" (NTDs) refers to a group of congenital malformations resulting from a defect in the closure of the neural tube. It occurs during the fourth week of embryonic development [1]. It is one of the most frequent congenital malformations in the world with an incidence rate of 5 to 10/1000) [2]. CT scan, transfrontanellar ultrasound and X-rays are the main examinations used to establish the diagnosis. In Madagascar, the

management of this anomaly faces therapeutic difficulties. The insufficiency of national data concerning this pathology justifies our study whose main objective is to identify the epidemiological and therapeutic profile as well as the short-term prognosis of Neural Tube Defects or NTDs.

Materials and Methods

This is a retrospective study carried out in the neurosurgery department of the CHUA-HJRA of Antananarivo over a period of 4 years from 01 January 2012 to 01 January 2016. Our study population consisted of patients who were hospitalized in the neurosurgery department for NTDs between 1 day of life and 34 months for neural tube closure anomalies. We used data from the hospital registry, patient medical records, paraclinical examination reports, and operative reports. The collected data were entered and analyzed using a PC with XLSAT version 2007 for Windows. This was done in order to highlight the incidence, the gender, the etiological factors encountered, the anatomical forms and finally the treatments that were performed.

Results

We collected 41 cases of NTD out of 1624 hospitalized pediatric population for a four year study, i.e. 2.5%. This gives an annual frequency of 10.25 cases per year and an incidence of 5 cases per 1000. The age of admission to the specialized structure varied between the first day and the first 3 months of life with a mean age of 14 days (Figure 1). A male predominance was noted with a sex ratio of 2. Prenatal discoveries could be made in 4.87%. The most frequent anatomical form was Meningocele (53.65%) followed by Myelomeningocele (36.5%, Table 1). Hydrocephalus was the most associated malformation and represented 26.82% (Table 2). Children born to mothers between 19 and 22 years of age were the most affected with a percentage of 34.14%. The average maternal age was 26.024 years (Figure 2). The majority of the mothers were multigestational for a percentage of 48.78%. In 51.21% of the cases, the mothers completed their prenatal consultations, while 34.14% were lost to follow-up and 14.63% did not do so. Folic acid intake during pregnancy was noted in 82.93% of women. In the whole series, we found 24 cases (58.53%) with the notion of fever during the first trimester of pregnancy. Five percent (5%) of the mothers had taken an anti-epileptic treatment during their pregnancy. No family cases were identified. Thirteen (13) patients (31.70%) presented with motor deficits of the lower limbs and abnormalities in their archaic reflexes. Sphincter genital disorders were observed in 9.75%. Only 03 patients presented trophic disorders (7.31%) (Table 2).

Figure 1: Distribution of patients according to age of care.

Anatomopathological aspect	Effective	Percentage (%)
Spina bifida occulta	01	2.43
Meningocele	22	53.65
Encephalocele	03	7.31
Myelomeningocele	15	36.5
Total	41	100

Table 1: Distribution of NTDs according to anatomopathological form.

Existence of associated malformations	Number	Percentage (%)
Orofacials	01	2.24
Heart defect	01	2.24
Digestives	04	9.71
Lung	01	2,24
Urological	01	2.24
Hydrocephalus	11	26.82
None	21	51.21
Total	41	100

Table 2: Distribution of NTDs according to associated malformations.

Figure 2: Clinical aspect of lumbar meningocele (CHUA-HJRA-Neurosurgery Department).

Figure 4: Distribution of patients according to the location of malformation.

The majority of the anomalies were located at the ethmoido-nasal level and at the vertebral column with a percentage of 41.46% and 36.58% respectively (Figure 3). Eleven cases of rupture of the malformations (26.8%) were noted. CT scans were systematically performed in all patients with NTDs for a more precise assessment of the lesions (Figure 4). Twenty-seven patients (65.85%) had this examination. In the absence of CT scan, 06 patients received an ultrasound scan and 07 patients received a standard X-ray.

On the therapeutic side, thirty-six patients (87.80%) underwent surgery to correct the malformation. Of these, 32 patients (81%) had a complete repair of their malformation and 04 patients (11%) had an incomplete removal. Postoperatively, 07 patients or 19.44% developed a CSF infection. Of the 41 cases of NTD observed we had 02 cases of death or 4.87% related to severe cardiopulmonary malformation. The appearance of certain neurological deficits, trophic and sphincter disorders concerned 17.83% of patients.

Figure 3: Distribution of malformations according to the maternal age.

Discussion

The number of patients hospitalized in the Neurosurgery Department for neural tube defects during the period of our study was 41 cases out of 7387 hospitalizations, i.e. an incidence of 5 cases per 1000. This gives an annual incidence of 10.25 cases per year. Studies done in Africa on spina bifida have shown similar annual frequencies [3,4]. On the other hand, in the developed countries of the West, frequencies ranging from 0.66 to 1 per 1000 births and 4 per 1000 live births are noted [5,6]. Thus, a high frequency in developing countries could be explained by the delay in advancing on prenatal diagnosis to indicate therapeutic termination of pregnancy. A low level or even absence of folic acid intake during the periconceptual period is one of the causes [2]. The age of care in our series varied between the first day of life and 3 months, with an average age of 14.5609 days for a percentage of discovery of 68.29% from birth. These results show that the vast majority of patients are seen early enough, i.e. within 72 hours of birth. The study by Coop reached the same conclusion [8]. The delay in consultation compromises the principle of early intervention suggested by Lorber J. [9] and exposes patients to complications. This late consultation could be explained by the distance from the major

specialized centers, and the fact that some parents are unaware of some aspects of the malformation.

The 41 patients were divided into 14 girls (34.14%) and 27 boys (65.85%). The sex ratio M/F was 2. Some authors have found a similar predominance [10,11].

Ante-natal diagnosis of the malformation is important to reduce the morbidity and mortality associated with it [3,12]. In our observation, only two cases of antenatal discovery were noticed. The study of G. Macé describes that the sensitivity of ultrasound varies according to gestational age and depends on the operator and the definition obtained with the apparatus used and the author concludes that the examination of the spine must be systematically carried out at the ultrasound of 22 SA. In case of difficulty in examining the fetal spine with anomaly associated with a high AFP rate, a much deeper examination will be performed by a specialized sonographer [5].

The irregular and non-existent prenatal consultations among some pregnant women, the lack of equipment in rural health centers and the lack of qualified personnel in some regions are at the origin of the delay in care.

In our study, we observed 53.65% of meningocele, 36.5% of myelomeningocele, 2 cases of lipomeningocele and no case of lipomyelomeningocele. It is thus essential to underline the importance and the place of medical imaging, particularly MRI, in the distinction and anatomical classification of this malformation.

In our series, hydrocephalus was the most common malformation found with a percentage of 26.82%. This result is almost similar to the 30.74% found by Sanoussi S [3], the 34.12% of Kabré A [8] and the 24% of Alhassane T in Mali.

No orthopedic malformations such as equine clubfoot were found, but anorectal malformations accounted for almost 9.71%. These results are almost similar to those found in Mali [10]. Fourteen of the 41 cases had no associated malformations. Thus, in daily practice, it is incumbent on health care personnel to perform a thorough and complete somatic examination, in order to detect possible malformations or associated anomalies; thus, allowing early management, while avoiding or preventing complications.

Concerning the age of the mother, in our study, the age group most concerned was that between 19 and 22 years and represented a percentage of 34.14%.

This result is in agreement with those of the literature where generally the extreme ages of 19 to 35 years are those where the predominance of NTDs is most found [14].

The majority of the mothers were multigestational in 48.78%. This risk is consistent with the literature where it is noted that neural tube closure anomalies increase with the number of pregnancies and deliveries [15].

Only 51.21% of mothers had completed their prenatal visits. Thirty-four percent of the mothers had not completely achieved their prenatal visits and 14.63% had not performed any prenatal visits. Our results are not consistent with the literature. According to Oumar B [16] and N'Diaye [17], the majority of mothers did not attend prenatal consultations, whereas 95% do in developed countries [18].

Concerning the intake of folic acid during pregnancy in the context of prenatal consultations, it was noted that 82.93% of women took folic acid during their pregnancy.

In 58.53%, i.e. 24 cases, we found the notion of fever during pregnancy without being able to pinpoint the exact period of this febrile attack. In 95.12% of cases, no mother had taken antiepileptic treatment during her pregnancy. Our study did not find any history of NTD in the siblings or in either parent. According to Lorber J [9], the risk of recurrence in the same family is estimated at about 3% (between 2 and 5 depending on the study) after the birth of an affected child, between 3 and 10% after 2 affected children, 12% after 3 affected children and 25% after 4 affected children.

At the time of admission of the patients, the general health condition was preserved in 63.34% of the patients, i.e. 26 cases, as opposed to 15 cases with an altered general health condition.

These results are similar to a study done in Mali by Sidi S. where out of the 62 cases studied, 15 of them presented an alteration of the general health state [19].

Thirteen patients (13) out of the 41 patients in our study, i.e. (31.70%) had a motor deficit of the lower limbs and abnormalities in their archaic reflexes and 68.29% had a normal neurological state.

In India, Kumar R., *et al*. [20], reported a higher result than ours, i.e. 66.5% of the patients had a motor deficit. Only 03 patients had trophic disorders (7.31%) and 38 cases or 92.68% had no disorders. Neural tube defects require multidisciplinary participation to ensure perfect urological, neurological and psychological management.

Thus, to do this, the availability of a satisfactory technical platform is essential. Our study allowed us to observe that in the majority of cases, the location of the malformation was at the ethmoidonasal level, with 17 cases observed, i.e. a percentage of 41.46%. The second most observed location was at the level of the spine with a representative percentage of 36.58%.

Our results are not similar to those of Kabré A [8] who noted 94.62% of caudal locations with, in order of frequency, lumbosacral, lumbar, sacral, dorsolumbar, cervical and dorsal locations.

CT was the basic complementary examination in our study. A cerebral CT scan was systematically prescribed to all patients for a precise diagnosis of neural tube defects. Transfrontanellar ultrasound was prescribed in some infants in the absence of a brain scan. 06 patients (14.63%) underwent this examination, thus confirming the diagnosis and highlighting the existence of associated hydrocephalus in some patients. In a study carried out by Sidi S [19] on 62 patients, 22 patients (35.5%) were able to perform a transfontanellar ultrasound. The others who were not able to perform this examination lacked financial resources.

We were able to consult spinal X-rays in some patients; 17.07% of the patients, i.e. 07 patients, had recourse to this examination. [19] in a study carried out in Mali was able to identify 18 patients (29.1%) who had undergone a spinal radiography. The 44 others who were unable to perform this examination lacked the means to do so. Since its advent, magnetic resonance imaging (MRI) has been considered the best means of exploring vertebro-medullary and cerebral dysraphic lesions [21].

This examination, in addition to being efficient in the assessment of soft tissue lesions, is also harmless for the patients. However, it is not available in our context. In order to have complete lesion assessments to better manage cases of NTDs in our context, we have highlighted the fact that in 39.02% of the cases the delay in management was between the first day of life and the third month of life and represented 16 patients.

The second most observed age group was between 1 and 2 years with a percentage of 24.39%. The average age of care was 9.96 months.

In general, this delay in intervention is related to the delay in diagnosis, relative contraindications to anesthesia such as: low birth weight, prematurity, intercurrent infections and financial problems, given that most children with NTDs come from underprivileged areas.

In our study of 41 cases, 36 patients, or 87.80 percent, underwent surgery to correct the malformation. This rate is close to that reported by Alatis [8] (60.19%) and Sanoussi S [3], which was 49.35% of patients operated. In our hospital, the main reason why some patients did not benefit from surgical treatment was the low socio-economic level and a lack of financial resources for management.

Concerning the evolution, 63.41% of the patients had a good evolution of clinical and paraclinical signs. Nine patients (21.95%) had a stationary evolution and in 14.63% there were recurrences. Seven patients (19.44%) developed a CSF infection after surgery. A study conducted by Sanoussi S., *et al*. [3] in their series showed a superficial infection of the surgical scar in 37.69 sometimes complicated by CSF infection.

We had 02 cases of death, i.e. 4.87% of patients with this pathology.

Ouattara O. in Ivory Coast [4] obtained 13 deaths/80 cases with a percentage of death of 16.25%. However, the multidisciplinary management of these children by a pediatrician, obstetrician-gynecologist, radiologist, neurosurgeon, and biologist has led to a considerable improvement in their condition. The major handicaps remain motor disorders and sphincter incontinence [22].

Eleven patients (30.55%) had persistence or appearance of neurological deficits, trophic and sphincter disorders a few weeks after their surgery. Nevertheless, the long-term follow-up after dis-

charge from hospital was difficult. This is due to the fact that after the surgical treatment the follow-up protocol is poorly respected by the patients. Many patients who are discharged do not return for follow-up consultations and are lost to follow-up.

Conclusion

This study carried out in the neurosurgery department of the CHU - HJRA allowed us to identify the epidemiological profile and the etiological factors of NTDs. The predominance of males was demonstrated in our study. There are factors related to the occurrence of neural tube defects such as multiparity, young age, a high maternal age, a notion of infection during pregnancy and the absence of folic acid intake. She also reminded us that the anatomoclinic forms are dominated by Meningocele and Myelomeningocele. The CT scan was the best and first confirmatory imaging test available to us. The main treatment was neurosurgical. The most common complications were CSF infection and hydrocephalus. This prognosis during our study was sometimes difficult to establish because of the loss of sight of the patients and the insufficiency of technical platforms. Folic acid intake is recommended for the prevention of NTDs. The realization of antenatal diagnosis through the practice of ultrasound could lead to early discovery of cases of neural tube defects. The availability of adequate technical facilities in the health centers remains one of the key points in the process of improving the management and follow-up of these diseases.

Periconceptional folic acid intake, adequate treatment of maternal infection, and systematic screening for NTDs during obstetrical ultrasound constitute the minimum and mandatory prescriptions during the prenatal consultation.

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Volume 4 Issue 10 October 2021