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CASE STUDY

The Reality of the Management of Spinal Dysraphism in a Country in a Precarious Situation: A Retrospective Study

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ABSTRACT

Background: **To report the epidemiological, therapeutic and evolutionary aspects of spinal dysraphism, with emphasis on the difficulties encountered during management in Madagascar.**

Methods: **This is a retrospective, descriptive, multicenter study in the 7 neurosurgical centers of Madagascar over 5 years on children operated on for spinal dysraphism.**

Results: **We collected 50 cases over 5 years. The mean age was 16.95 ± 33.94 months. Girls predominated (sex ratio = 0.85). The absence of folic acid supplementation in the mother was found in 62%. In all cases, the diagnosis was made in the postnatal period. Myelomeningocele is the most frequent form. Hydrocephalus was found in 20% of cases, orthopaedic malformation in 20%, and Chiari type II malformation in 2%. The dysraphism was localized at the lumbar level in 50% of cases. A spinal CT scan was performed in 62% of cases and transfontanellar ultrasound in 22% of cases. No magnetic resonance imaging of the brain or spinal cord was performed. A ventriculoperitoneal shunt was performed in 8% of cases. Postoperative hydrocephalus was 4% and mortality was 1% due to meningitis. The average follow-up time was 3 months with an extreme of 3 months to 3 years.**

Conclusion: **In spite of the difficulties in the management of these children and the technical platform available in the country, they are operated correctly and the hospital mortality is low.**

Keywords: Infant, Spinal dysraphism, Tomography.

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1. Introduction

Spinal dysraphism is a rare congenital malformation secondary to a neural tube closure defect occurring between the 22nd and 28th day of pregnancy. This malformation can lead to a major functional handicap because of the motor and bladder-sphincter disorders it causes. It can also cause a mental disorder secondary to the associated hydrocephalus. In France and the United States, the total prevalence is 1 per 500 births [1], [2]. The objective of our study is to report the epidemiological, therapeutic and evolutionary aspects of spinal dysraphism, with emphasis on the difficulties encountered during management in Madagascar.

2. Materials and Methods

This is a retrospective, descriptive, multicenter study in the 7 Neurosurgery centers of Madagascar during 5 years from January 01, 2017, to December 31, 2021. We included all children operated for spinal dysraphism managed in these services.

The parameters studied were:

- Incidence, age, sex, geographical origin and history, risk factor;
- Mode of discovery of the malformation, topography, skin aspect, associated malformations and neurological signs;
- Imaging studies performed (type of malformation, presence or absence of associated hydrocephalus/Chiari type II malformation);

- Age of the children at the time of surgery, type of surgery;
- Postoperative complications (hydrocephalus, CSF leakage through the scar);
- Mortality rate;
- Duration of follow-up.

Several documentary sources were used to carry out this study:

1. The departmental inpatient registry, which lists all patients hospitalized in the department (name, age, telephone number, medical record number, reason for admission, and discharge diagnosis).
2. Patient medical records, which contain each patient's personal data, detailed medical observations, treatment and follow-up records, and results of additional tests performed.
3. The surgical notebook, which contains the name and age of the patients, the name of the surgeon, the type of operation, the course of the operation and a clinical summary. The collected data were entered into Microsoft Excel 2019® and analyzed on R® software version 3.5.2 with an IDE (Integrated Development Environment) RStudio® version 1.1.456.

Prior to data collection, permission was obtained from the relevant hospital directors and department heads—respect for anonymity, confidentiality, and personal and professional secrecy. The forms containing the information collected are kept in a safe place.

3. Results

We collected 50 cases in 5 years, i.e., 10 cases per year. The majority of cases came from the capital (Antananarivo) in 56% of the cases and from Mahajanga (west of the country) in 30% of the cases. The mean age was 16.95 ± 33.94 months. Women predominated (sex ratio = 0.85). The absence of folic acid supplementation in the mother was found in 62%. The intake of fenugreek grains by the mother was recorded in 33% of cases in Mahajanga. In all cases, the diagnosis was made after birth.

Myelomeningocele (50%) was the most frequent form, followed by meningocele (40%) and then spina-lipoma (10%), and no cases of occult dysraphism without subcutaneous mass were diagnosed. Hydrocephalus was found in 20% of cases and orthopedic malformation in 20% of cases. Chiari type II malformation was found in 2% of cases (Table 1). The dysraphism was localized in the lumbar region in the majority of cases (Fig. 1). Motor deficit was found in 8% of cases and sphincter disorder in 12%. The pocket was infected in 4% of cases.

A spinal CT scan was performed in 62% of cases and a transfontanellar ultrasound in 22% of cases. No MRI of the brain or spinal cord was performed. The age of the children at the time of surgery was 17 months, a repair was performed in all cases, and a ventriculoperitoneal shunt was performed in 8% of cases. Postoperative hydrocephalus was 4%, and cerebrospinal fluid leak at the scar was 4%. The mortality rate was 1% due to meningitis. The average follow-up time was 3 months with an extreme of 3 months to 3 years.

TABLE I: Distribution of Children by Associated Malformations

Associated anomaly	Number (n = 50)	Percentage (%)
Hydrocephalus	10	20
Orthopaedic malformation	10	20
Atrial septal defect + persistence of the arterial canal	1	2
Chiari malformation type II + Inguinal hernia	1	2
Umbilical hernia	1	2
No associated malformation	27	54

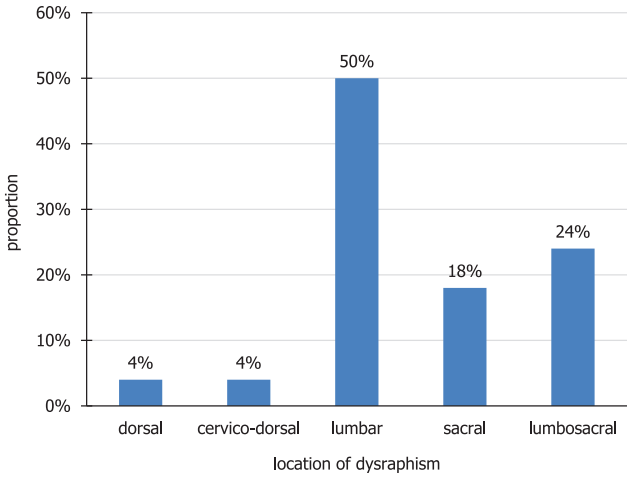

Fig. 1. Distribution by location of dysraphism.

4. Discussion

Due to the lack of CT scanners in the provinces, patients are obliged to come to the capital to benefit from them and prefer to stay in Antananarivo for the operation in order to avoid making round trips. This is why the majority of cases have been reported in the capital.

After Antananarivo, Mahajanga is the second most frequent source of spinal dysraphism, because in the western regions of the country, pregnant women tend to take fegnegrec, which increases the risk of developing a neural tube defect according to Moroccan studies [3].

Literature studies have shown that proper maternal folic acid intake reduces the risk of dysraphism by 70%. Periconceptional supplementation is recommended at best 4 weeks before the start of pregnancy and up to 12 weeks of pregnancy. The recommended dose is 0.4 mg/day for the general population. It is increased to 5 mg/day for women with a history of a child with AFTN, pre-existing diabetes and anti-epileptic treatment [4]. Therefore, we need to reinforce the IEC on the importance of folic acid supplementation in our country, as 62% of mothers have not been supplemented.

In developed countries, the diagnosis of spinal dysraphism is made prenatally in more than 85% of cases of myelomeningocele. In these countries, in-utero surgery is performed in some cases of myelomeningocele, and medical termination of pregnancy is also proposed for myelomeningocele whose prognosis meets the requirement of a particular severity, even in case of in-utero surgery [5], [6]. This is not yet possible in our country. Our result

confirms the data in the literature concerning the predominance of myelomeningocele for spinal dysraphism, which is the most severe form. Currently, there is no universally accepted classification for spinal dysraphism [2], [7]. However, the most widely used classification at present is that of Thompson *et al*. [2]. They classified dysraphism not only according to the presence or absence of skin coverage but also according to the embryological basis of the dysraphism. Open dysraphism is characterized by exposure of nerve and meningeal structures without skin coverage: myelocele exposure of the spinal cord and/or root without meninges and myelomeningocele exposure of the spinal cord and root with meninges. Closed dysraphism with subcutaneous mass is represented mainly by meningoceles and spina lipoma and closed dysraphism without subcutaneous mass. The latter form is often revealed by the presence of a skin stigma, the dermal sinus is the form that requires urgent management [2]. In our series, no cases of closed dysraphism without subcutaneous mass were diagnosed.

In developed countries, MRI of the brain and spinal cord is routinely performed in the first days of life for open dysraphism, and MRI of the spinal cord at 2 to 3 months of age for closed dysraphism. A transfontanellar ultrasound is systematically performed at birth to measure the size of the ventricles and to allow the follow-up of a possible hydrocephalus [8], [9]. In Madagascar, access to MRI is still difficult because of its high cost, so spinal CT and transfontanellar ultrasound are most often requested.

We found associated hydrocephalus in 10% and Chiari type II malformation in 2%, whereas according to the literature in 80% of cases, dysraphism is associated with hydrocephalus and Chiari II and in 100% of cases for myelomeningel but hydrocephalus is not always surgical [1], [4]. In Madagascar, due to a lack of resources, systematic brain imaging is not possible for cases of myelomeninges and therefore hydrocephalus and Chiari type II malformation are rarely diagnosed.

In our series, the age of the children at the time of surgery was 17 months. In developed countries, surgery is performed in the first days of life for open dysraphism. It is performed in the first month of life for closed dysraphism. A ventriculoperitoneal shunt is performed in case of hydrocephalus with intracranial hypertension or in case of progressive hydrocephalus [1], [9]. We have seen a delay in treatment compared to developed countries. This is because the majority of parents first bring their children to traditional practitioners and only when there is no improvement do they go to the hospital.

The average length of follow-up is 3 months, so the majority of children are lost to follow-up after the first visit. The outcome of these children in adolescence and adulthood is not known, especially with regard to schooling and independence.

5. Conclusion

The main risk factor is the lack of folic acid supplementation in pregnant women. Meningoencephalitis is the most common form and is the most severe form. Access to imaging is difficult, and a delay in management has been noted. The majority of children are lost to follow-up during the first consultation. Despite these difficulties and the technical facilities available in the country, these children are operated on correctly and hospital mortality is low. A multidisciplinary team composed of neurosurgeons, orthopedists, traumatologists, urologists, obstetricians, and psychologists, with the development of a national protocol of diagnosis and care and a national center of reference with secure funding for these children, could improve the management. A prospective study is necessary to know the long-term future of these children.

5.1. *What Do We Know About this Subject?*

- The main risk factor is the lack of folic acid supplementation in pregnant women.
- Myelomeningocele is the most frequent form and is almost always associated with hydrocephalus.
- In developed countries, surgery is performed in the first days of life for open dysraphism. It is performed in the first month of life for closed dysraphism.

5.2. *At this Study Adds*

- Despite these difficulties and the technical facilities available in the country, these children are operated on correctly and hospital mortality is low.
- A multidisciplinary team composed of neurosurgeons, orthopedists, traumatologists, urologists, obstetricians, and psychologists, with the development of a national protocol of diagnosis and care and a national center of reference with secure funding for these children, could improve the management.

Limitations

This is a retrospective study, and therefore subject to the effects of confounding factors. The sample size is small.

Author Contributions

Radotina Tony Andrianaivo, Willy Francis Rakotondraibe, Bemora Joseph Synese: substantial contributions to conception and design, acquisition of data, or analysis and interpretation of data; drafting the article or revising it critically for important intellectual content. Willy Ratovondrainy, Mamiarisoa Rabarijaona, Clément Andriamamonjy: final approval of the version to be published.

Conflict of Interest

Authors declare they do not have any conflict of interest.

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