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Original Article

The prevalence and pattern of neural tube defects and other major congenital malformations of nervous system detected at birth in Barbados

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ABSTRACT

Introduction: Neural tube defects are the commonest major congenital malformation of nervous system. There is paucity of published data on the public health burden of major congenital malformations from the English Caribbean. The objective of this study was to describe the prevalence and pattern of major congenital malformations of the nervous system seen at birth. We also describe the burden of neonatal morbidity and mortality from the neural defects.

Methods: This was a retrospective clinical audit of all babies born with major congenital malformations of the nervous system in Barbados. The period of reporting was from 1993 to 2012. Relevant data on the babies with major congenital malformations of the nervous system were obtained from the birth register in the labor ward and the admissions register at the NICU.

Result: The prevalence rate for neural tube defects, congenital hydrocephalus, and other Major Congenital Malformations of the Nervous System was 3.76, 2.66, and 1.25 per 10,000 live births, respectively. Major congenital malformations of nervous system accounted for 10 (1.6%; 95% CI = 0.8%–3.1%) deaths. The prevalence rate of neural tube defects is lower than those reported from most of the countries around the world.

1. Introduction

Major congenital malformations of the nervous system (MCMNS) are reported to be one of the most common major congenital malformations in newborns.[1–4] The prevalence rate of the MCMNS in reports from Asia, Middle-East, Africa, and Europe varies between 23 and 61 per 10,000 live births.[5–8] There is a wide variation in the prevalence of these malformations in different parts of the world.[9] MCMNS are an important cause of mortality and long-term morbidities among children.[8–11] They are becoming relatively more common cause of long-term morbidities and mortalities among children due to the decreasing morbidities and mortalities from causes such as perinatal injuries, infections, and corrective surgeries for most congenital heart diseases. Historically, perinatal injuries, and congenital heart diseases were the common causes of long-term morbidity and mortality among children. Long-term and often disabling morbidities from the malformations of the nervous system poses a serious economic and social burden on the government and the society.[11–13] Many of these malformations of the nervous system such as the neural tube defects, which is the most common of these malformations, can be prevented.[14–17] In others, antenatal detection can help to reduce prevalence, long-term morbidities and mortalities.[16,18]

Barbados, one of the English speaking Caribbean countries, has a total population of 283,000 (2012) including 58,500 (21%) and children under the age of 16. The Under 5 Mortality Rate was 18 per 1000 live births and the life expectancy at birth male/female was 75/81 years.[19] It has a well-organized state run health care infrastructure with free health care for its citizens at the point of delivery.[20] Over 95% of pregnant women having had at least two antenatal visits and over 90% of all deliveries in this country take place at the Queen Elizabeth Hospital.[21] Over one-eighth of all neonatal death in this country is attributed to the major congenital malformations.[22] In this study, we describe the overall prevalence rate and the long-term trends for the MCMNS detected at birth in this population. We also describe the pattern of nervous system

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malformation, the factors associated with these malformations, and the impact of these malformations on the overall neonatal mortality rate in this country.

2. Material and methods

This is a clinical audit of babies born with MCMNS from among all the deliveries in Barbados. The period of audit extends from 1993 to 2012. The stillbirths with MCMNS were excluded from this study. Babies with diagnosis of any of the MCMNS were identified from the admission register maintained in the Neonatal Intensive Care Unit (NICU) at the Queen Elizabeth Hospital (QEH). Relevant data on the babies with MCMCNS were obtained from the birth register in the labor ward and the admissions register at the NICU. These included date of delivery, maternal age, maternal parity, gestational age at delivery, birth weight, and baby's gender. The total number of live births, number of neonatal admissions, total number of babies born with major congenital malformations, death among babies with any major malformations and number of deaths in babies with MCMNS, and overall total number of neonatal deaths during the study period were also collected form these registers. The hospital case records for all the babies with MCMNS were also reviewed to collect additional maternal data such as history of trauma, infection, chronic diseases, medications including over the counter preparations, use of alcohol or drugs in pregnancy. The baby's data such as other associated malformations and outcome in terms of hospital discharge or death were also collected from the baby's case record. Birth statistics for the minority of the births outside the hospital were also collected.

As a routine practice all live born babies were examined after birth by a pediatrician and congenital anomalies, if any, were described in detail. All babies with malformations were appropriately investigated with neuroimaging including ultrasound examination of brain and CT scan of brain. MRI scan and genetic studies were unavailable during much of the study period. Neonatal treatment data and medical complications as recorded by the attending neonatologists at the time of death or discharge from the hospital were also collected.

Throughout the study period discharge/death diagnosis was recorded using the clinical approach for the classification of the major malformations of the nervous system. Major congenital malformations included defects present at birth, which may be fatal and/or handicap the patient throughout life. Based on the review of the neuroimaging studies and other investigations we classified these malformations based on those proposed by Van der Knapp.[23] Prevalence was calculated by dividing the number of newborns with the MCMNS (numerator) by the total number of live newborns for the given time period (denominator) and expressed as per 10,000 live births. Proportion of neonatal deaths attributed to MCMNS was calculated by dividing the number of death attributed to MCMNS during the first month of life by the total number of all deaths during the first 28 days of life expressed as percentage. Case fatality rate for the MCMNS was calculated by dividing the number of deaths during the first 28 days of life by the total number of live born babies with MCMNS requiring admission to the NICU. Prematurity was defined as: delivery less than 37 complete weeks of gestation. Small for Gestational Age (SGA) was defined as the birth weight below the 10th percentile for the given gestational age.

Ethical approval was obtained from the ethics committee at the Queen Elizabeth Hospital and Institutional Review Board for ethics in study involving human subjects of the University of the West Indies and the Ministry of Health, Barbados. All precautions were taken to protect the personal information of the patients, only the investigators have access to the database.

Data were entered and stored in a specially created Microsoft Access spreadsheet, Microsoft Excel was used for data tabulation and generation of graphs. The results were analyzed by simple statistical techniques and tests of significance including Chi-square tests were applied. The online statistical calculator Vassar Stats was used for calculation of 95% Confidence Intervals (CI) with continuity and the Relative Risk (RR).

3. Results

Over the 20 years study period, there were 63,827 live births in Barbados and there were 49 cases of MCMNS. The overall prevalence rate for the MCMNS during the 20 years study period was 7.68 per 10,000 live births. The relative proportion of the main category of MCMNS based on the discharge/death diagnosis is shown in Fig. 1. Neural tube defects with or without hydrocephalus were the commonest (49%; 95% CI = 34.6–63.5%) malformation seen. It was followed by congenital hydrocephalus (34.7%; 95% CI = 22.1–49.7%) not associated with neural tube defects. Congenital hydrocephalus not associated with neural tube defects consisted of Chiari Malformations (13 cases), Dandy Walker malformation (2 cases), and Congenital hydrocephalus not associated with any other malformation on CT scan (2 cases). The prevalence rate for the neural tube defects and the congenital hydrocephalus was 3.76 and 2.66 per 10,000 live births, respectively. Trend in the prevalence rates of various types of MCMNS are shown in Fig. 2. The overall prevalence rate for all MCMNS increased from 6.99 per 10,000 live births during the 1993–2002 to 8.40 per 10,000 live births during the 2003–2012. However, this difference in the prevalence rate over the time period was statistically not significant (RR = 1.21 and $P = 0.512$). Over the same time period, the prevalence rate for the neural tube defects increased from 3.47 to 4.04 per 10,000 live births. This difference in the prevalence rate too was statistically not significant ($P = 0.708$). The risk ratio for the neural tube defect during the latter ten-year period compared to former ten year period was 1.16 (95% CI = 0.52–1.16).

The details of the pattern of MCMNS seen in this population, based on the Van der Knapp classification system, are shown in Table 1. Disorder of dorsal induction resulted in 75.5% (95% CI = 60.8–86.2%) of all MCMNS, with neural tube defects accounting for 49% (95% CI = 34.6–63.5%) of all MCMNS. Myelomeningocele was the commonest (62.5%; 95% CI = 40.8–80.4%) of the neural tube defects, accounting for 30.6% (95% CI = 18.7–45.6%) of all MCMNS. Of the 15 cases of Myelomeningocele, hydrocephalus, and Chiari type 1 malformation was detected on CT scan of brain in 10 (66.7%; 95% CI = 38.7–87.0%). Anencephaly was the least common

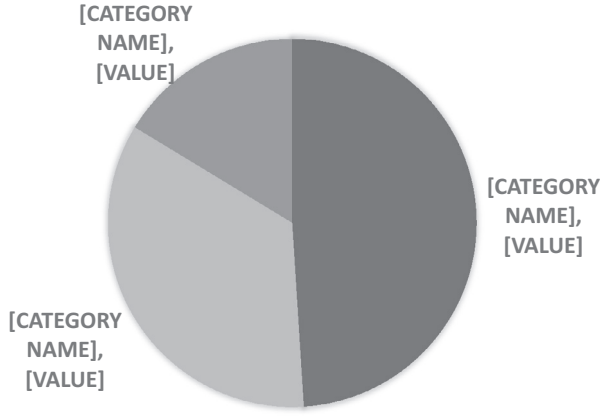

Fig. 1. Distribution of the clinical diagnosis categories of the major congenital malformations of the nervous system in Barbados, 1993–2012.

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