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A Case of a Ruptured Meningomyelocele: A Plastic Surgeon's Approach in Jamaica

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Abstract

Meningomyelocele (MMC) is one of the adverse birth outcomes in Jamaica related to a low socioeconomic status and health care limitations. We present a case of a male neonate with a ruptured lumbar MMC, hydrocephalus, Chiari type 2 malformation and talipes equinovarus. Surgical repair of the MCC accompanied by reconstruction of the skin defect with a transposition flap was accomplished within 48 hours. Subsequently there was the insertion of a ventriculoperitoneal shunt. The reconstructive surgeon is posed with a challenge to select the ideal method of closure that is tailored to the shape and size of this large defect while working in a multidisciplinary team.

Keywords: *Meningomyelocele Folic acid, Neural tube defects Rhombic flap.*

Introduction

Meningomyelocele (MMC) is a severe type of spina bifida cystica and the commonest congenital malformation of the central nervous system (CNS) compatible with life [1,2]. However, in untreated patients the mortality rate in the first 6 months of life is 35% - 70% [3, 4]. There is cystic herniation of meningeal and spinal cord tissue outward through the defective bony arches and skin as a sac [3,4]. It may be covered by skin or a thin, easily ruptured membrane and may occur anywhere along the vertebral column [1]. However, they are the most common in the lumbosacral region [1,3,4]. Although the exact aetiology of MMC is unknown, it could be multifactorial, including genetic, racial, environmental factors, geography, nutrition, gestational diabetes, maternal obesity, low socioeconomic status, exposure to high temperatures early in pregnancy and some antiepileptic drugs [5-8]. Compelling scientific evidence has linked increased intake of folic acid during the periconceptional period with the prevention of neural tube defects (NTDs) [1,2, 5,6,7, 9,10].

Despite the fact that MMC is a potentially preventable cause of perinatal morbidity and mortality, it continues to be a prevalent disease among the low-socio-economic population in the Caribbean [3,11]. Jamaica has a high incidence of adverse birth outcomes related to several health care constraints that affect timely access to maternal care. Poverty is a contributing factor and to illustrate the point, the overall poverty rate in the island of 19.9% in 2010, increased by a further 2.3% by 2012 [12]. The last quoted incidence that we found of live-birth NTDs in Jamaica was the approximated period of 1978-1988 of 1.4 per 10,000 live births which then increased to 5.0 per 10,000 in one quarter due to the impact of Hurricane Gilbert [9]. The global incidence quoted in the literature ranges from 0.8 - 1 per 1000 live births [3, 6].

The disease imposes a high cost of treatment and burden to the family due to life-long disabilities such as paraplegia, incontinence, hydrocephalus, cardiovascular anomalies and mental retardation [3,5].

Approximately 10 cases of sacral MMC are managed by The Bustamante Hospital for Children, Kingston - The Regional Paediatric Institution in Jamaica - annually, usually within 24 - 48 hours of birth, as an emergency. This is generally in keeping with the literature with the exception of 2 sources allowing up to 72 hours post birth to commence surgery [2,4- 6].

A multidisciplinary team approach is warranted and the varying shapes and sizes of the defect pose individual challenges for the reconstructive surgeon in designing the ideal method for closure.

Materials and Methods

This is a retrospective study and literature review of a case report of a ruptured MMC at The Cornwall Regional Hospital, Montego Bay that was referred to The Neurosurgery Service for management immediately at birth.

History: The case involves a live term male neonate with a gestational age (GA) of $39^{+1}/40$ born before arrival to the hospital to a 30-year-old mother, gravida 4, para 4. There was no family history of NTDs or other congenital anomalies. The pregnancy was confirmed at an approximate GA of 12/40, however official antenatal care at the health centre was not sought until approximately GA of 17/40. The only investigation performed during pregnancy was an ultrasound done at this time, due to the absence of foetal movements, and no anomalies were reported. A diagnosis of anaemia in pregnancy presumed to be secondary to iron deficiency was made at a gestational age of $21^{+5}/40$ and she was placed on oral iron supplementation twice daily. Additionally, folic acid supplementation was commenced at an approximate GA of 20/40 and continued along with the iron supplementation until about a GA of 32/40. She was referred to the Cornwall Regional Hospital high risk antenatal clinic as a consequence of Caesarean- section 6 years previously, at a GA of 24/40. The remainder of the pregnancy was uneventful with a total of 8 antenatal visits.

The male neonate had Apgar scores of 7 and 8 at 1 and 5 minutes respectively. The weight was 7.63 lbs, length of 48.5 cm and with a head circumference (HC) of 33 cm. The significant examination findings were confined to the CNS, musculoskeletal (MSK) and dermatological systems. There was a bulging anterior fontanelle, distended scalp veins, splayed cranial suture lines, a ruptured lumbar MMC 4 cm x 4 cm with minimal cerebrospinal fluid (CSF) actively draining, a mongolian patch to lumbosacral region and talipes equinovarus. The neonate was commenced on antibiotics as a result of meconium-stained liquor. Cranial and abdominal ultrasound scans done at 1-day old revealed callosal dysgenesis with colpocephaly, a small posterior fossa with a likely Chiari malformation, likely germinolytic cysts and normal abdominal organs.

Surgeries: At 2 days of life there was a combined surgery with the Neurosurgery and Plastic Surgery teams for a duration of 6 hours. Closure of the MMC and a rhombic flap were completed under sterile conditions and with non-latex powder-free gloves. The flap was closed in 2 layers with 5.0 vicryl in the subcutaneous tissue and 5.0 prolene continuous interlocking sutures for the skin. Antibiotic ointment was applied, a gauze dressing and tegaderm applied. Care was taken to apply the dressings lightly with no pressure on the operative site. Postoperatively, the patient was nursed in a prone position with the head down and the pelvis raised. The antibiotic regime was continued.

At 25 days of life - postoperative day 23 – the rhombic flap was revised with excision of devitalized tissue at the upper portion and primary closure was achieved with subcutaneous and continuous with 4.0 monocryl skin sutures. The patient was placed on intravenous antibiotics for 5 days and there was a 2- hour turning schedule from the prone to lateral positions. At 41 days of life the neurosurgery team inserted a right ventriculo-peritoneal (VP) shunt.

Results/Observations

At 3 days postop, the tip of the flap became dusky with marked ecchymosis and obvious decreased perfusion over its distal 3cm. This was thought to be mainly limited to the epidermal layer, and was managed expectantly with copious antibiotic intment application. Over the next few days, the ecchymosis gradually subsided except for the distal 0.5 cm to 1 cm which progressed to full-thickness skin loss.

At 11 days postop a CSF leak was noted at the tip of the flap and this coincided with a progressive hydrocephalus and an increase in Head Circumference from 33cm at birth to 45.5 cm.

While the rhombic flap remained viable post revision, there was some delay in wound healing due to the CSF leak, which kept the suture line soaked. However, after placement of the VP shunt and elimination of the CSF leak, there was a rapid and complete healing of the wound.

The patient was subsequently discharged home at 44 days old- and 42 days post the first operation - and the flap has remained viable.

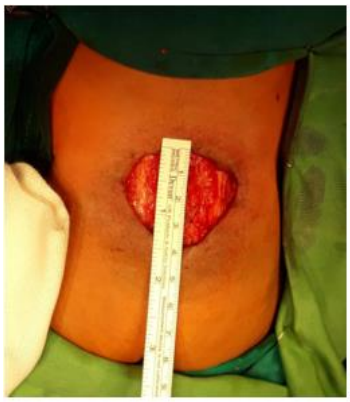

Figure 1: Skin defect post surgical closure of MMC.

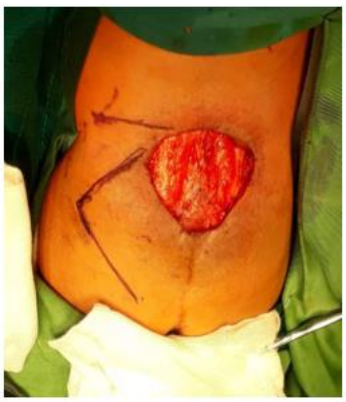

Figure 2: Surgical markings for raising the rhombic flap.

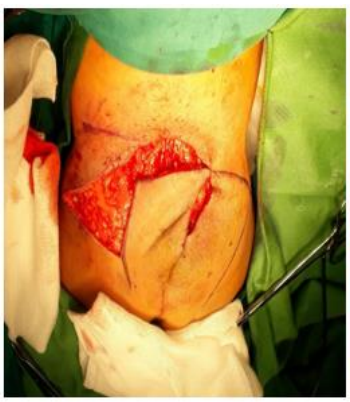

Figure 3: Rhombic flap has been raised and is being inset.

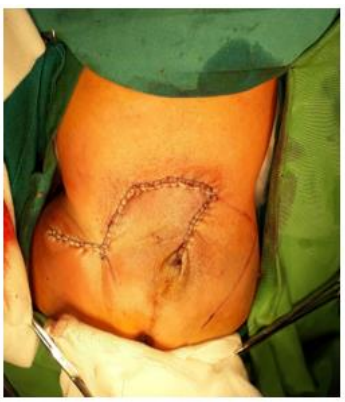

Figure 4: Closure of the defect with a rhombic flap.

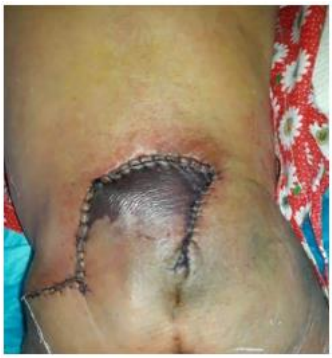

Figure 5: Postop day 3 after the first surgery showing ecchymosis.

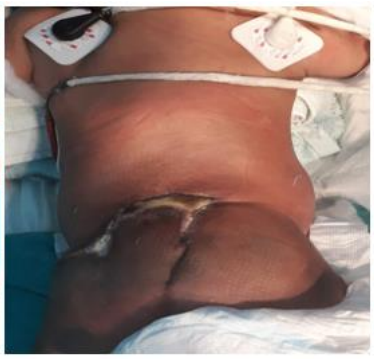

Figure 6: On the surgical table prior to the revision of the flap.

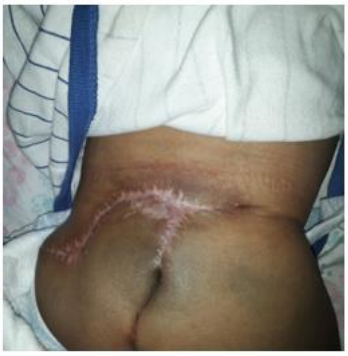

Figure 7: Completely healed, 3 months after the first closure.

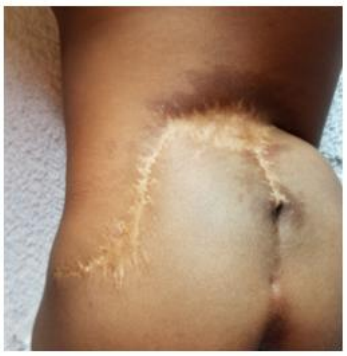

Figure 8: Three years post the first surgery.

Discussion

Factors associated with open neural tube defects the neural tube closes between days 17 and 30 of gestation [3]. The periconceptual period and the aforementioned range is the most critical for folic acid in prevention of these NTDs. Some women are unaware at this point that they may be pregnant as was the case with the mother. She did not commence folic acid supplements until 4 months into the pregnancy.

A report from Jamaica indicated that consumption of dietary folate during periconception was significantly lower among mothers of NTD cases than among matched controls as well the marked increase in NTDs post the major hurricane of 1988 [9,11]. Furthermore, antenatal care was not sought until at least a month after the confirmation of the pregnancy. This could be due to a possible combination of poor health-seeking behaviour in addition to a low socioeconomic status. Although it was documented that the dizziness in the current pregnancy was attributable to anaemia and likely secondary to iron deficiency, she also had the same complaint in the previous pregnancy 6 years previously and she may have been undernourished since that time. It is important to note that other nutritional deficiencies, aside from iron, impact anaemia and these include folic acid and vitamin B12. Additionally, parasitic intestinal helminth infection and bacterial infections which are present in Jamaica can contribute to anaemia [12]. A local study suggested the mandatory use of folic acid supplements and a fortification programme to be considered [11]. Along with the aforementioned, as Jamaica possesses a subset of those of a lower socioeconomic status to which the mother belongs, some excellent food sources to consider are lentils or any brown legumes, orange juice and home-grown green vegetables [9,12,13]. Although folic acid has its merits there must be caution with possible overdose as it can confound the diagnosis of pernicious anaemia due to vitamin B12 deficiency leading to irreversible neurological disorders and it may also be of concern in other diseases [10].

Early detection of neural tube defects

The fetal vertebral column can be detected by ultrasound at 10 to 12 weeks of gestation [1]. Although the ultrasound scan was done at 17 weeks in this case and revealed no anomalies, this may most likely be due to the operator-dependent results. The literature has stated the use of intrauterine fetal surgery for MMC repair with flaps which has been shown to result in improved neurological function, however they are not without risks [4,5,11,17]. This was not a feasible option in our setting, although it may have assisted in decreasing the severity of the Chiari 2 malformation and talipes equinovarus [11].

Reconstruction of a ruptured meningomyelocele skin defect

MMCs are usually closed within the first 24 - 48 hours of life, and our case was completed on the second day of life [5,6,11]. The aim of MMC reconstruction is to prevent ascending infection of the CNS and to provide durable skin and soft tissue coverage of the defect with minimal morbidity [4]. It should be considered as a surgical emergency requiring immediate closure. The technical difficulties are much less during the first 48 hours of life than they are at a later stage [4].

The vast majority, up to 75%, of the MMC skin defects are and can be reconstructed with direct closure while the remaining 25% representing large defects require other methods [2,5,11,15]. It has been documented since 1955 that for direct closure it is imperative that the undermining be carried out sufficiently in all directions to avoid tension on the suture lines and their subsequent dehiscence [16].

Proper preoperative planning and working in harmony with the Neurosurgery team are critical to reconstruction and will assist to shorten the operating time [5,8,11]. The MMC is assessed based on the size of the defect and whether there is a ruptured sac [5,11].

A recent study protocol stated that a defect with a total surface area of 18cm^2 and less could be closed by direct repair unless one of the diameters exceeded 4cm; a surface area of 18 - 80 cm^2 was closed by a rotational flap and greater than 80 cm^2 was closed by a skin graft.

The first mentioned use of the Limberg flap for closing MMC defects was by Ohtsuka et. al in 1979 [6,15]. A rhombic or Limberg flap was chosen in this case utilizing the available nearby skin laxity. However, there was limited redundant skin with which to work with, and after exploring various orientations of the flap, the best possible option was chosen to allow for primary closure of the donor area. In addition,

the thinness of the skin in this case was remarkable**,** and even with deepening of the dissection at its base, the flap was thin throughout and this was thought to be the cause of the circulatatory problems at its tip. The advantages of a rhombic flap in MMC defects are that they may be rapidly dissected, possess a relative ease of raising, can cover a round defect, have lower complication rates and preserve the back muscles. Several studies have documented the use of rhombic flaps with some dehiscence; however, all of the flaps went on to complete healing [5,6,15].

Wound site infection and CSF leak are the most common short-term complications of MMC repair [5]. The CSF leak in this case was primarily driven by hydrocephalus which accompanies MMC in nearly 80% of all cases [2,3]. Persistent CSF leakage is aborted by inserting a VP shunt in cases of progressive hydrocephalus [8] and resulted in rapid flap healing.

In conclusion MMC is to be considered a surgical emergency to be operated within 48 hours of birth. It requires a harmonious multidisciplinary team. The ideal approach is tailored with proper preoperative planning of a tension-free closure and meticulous postoperative care.

Informed Consent
Informed consent was obtained for the use of all the images in this report.

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Conflicts of Interests
None declared.

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