Myelomeningocele case series

In a retrospective analysis of CSF cultures of 45 MMC patients operated on during the years of 2002 to 2013. Before repairing the defect, the sac area was cleaned and three milliliters of CSF were drawn and sent for cerebrospinal fluid analysis for red blood cells, white blood cells, glucose level, protein level, chloride level, gram stain and culture. The CSF sample results were analyzed for irregularities in the values before proceeding with placement of a ventricular shunt.

All the CSF samples that were studied had at least 1 abnormal value in their results, even though none grew any pathogens in the cultures analyzed.

Upon CSF analysis, Mayol et al. found increased levels of cerebrospinal fluid proteins and other abnormal values in this population; however, none of the cultures grew any pathogens. This finding is an important tool in the evaluation of the possible etiologies of and therapeutic approaches for future shunt problems in this group of patients [1)].

2019

The aim of a study was to report the operative characteristics and outcome of a series of Iranian patients with large MMC defects utilizing the V-Y flap and with latissimus dorsi or gluteal muscle advancement.

This comparative study was conducted during a 4-year period from September 2013 to October 2017 in the pediatric neurosurgery department of Shiraz Namazi Hospital, Southern Iran.

The authors included 24 patients with large MMC defects who underwent surgery utilizing the bilateral V-Y flap and latissimus dorsi and gluteal muscle advancement. They also retrospectively included 19 patients with similar age, sex, and defect size who underwent surgery using the primary or delayed closure techniques at their center. At least 2 years of follow-up was conducted. The frequency of leakage, necrosis, dehiscence, systemic infection (sepsis, pneumonia), need for ventriculoperitoneal shunt insertion, and mortality was compared between the 2 groups.

The bilateral V-Y flap with muscle advancement was associated with a significantly longer operative duration ($p < 0.001$) than the primary closure group. Those undergoing bilateral V-Y flaps with muscle advancement had significantly lower rates of surgical site infection ($p = 0.038$), wound dehiscence ($p = 0.013$), and postoperative CSF leakage ($p = 0.030$) than those undergoing primary repair. The bilateral V-Y flap with muscle advancement was also associated with a lower mortality rate ($p = 0.038$; OR 5.09 [95% CI 1.12-23.1]) than primary closure. In patients undergoing bilateral V-Y flap and muscle advancement, a longer operative duration was significantly associated with mortality ($p = 0.008$). In addition, surgical site infection ($p = 0.032$), wound dehiscence ($p = 0.011$), and postoperative leakage ($p = 0.011$) were predictors of mortality. Neonatal sepsis ($p = 0.002$) and postoperative NEC ($p = 0.011$) were among other predictors of mortality in this group.

The bilateral V-Y flap with latissimus dorsi or gluteal advancement is a safe and effective surgical approach for covering large MMC defects and is associated with lower rates of surgical site infection, dehiscence, CSF leakage, and mortality. Further studies are required to elucidate the long-term outcomes [2)].

A prospective study of Zarutskie et al., from the Baylor College of Medicine, Texas Children's Hospital, Lucile Packard Children's Hospital Stanford from fetuses diagnosed with open neural tube defect that had in-utero myelomeningocele repair between April 2014 and April 2016. Independent variables were collected from four chronological sets of fetal images: pre-surgery ultrasound, pre-surgery MRI, 6-week post-surgery MRI and pre-delivery ultrasound. The following independent variables were collected from all image sets unless otherwise noted: gestational age, head circumference, mean ventricular width, ventricular volume (VV, MRI only), hindbrain herniation (HBH) score (MRI only), and level of lesion, defined as the upper bony spinal defect (pre-surgery US). Based on these measurements, additional variables were defined and calculated including change in degree of HBH, ventricular width growth (mm/week), and ventricular volume growth (ml/week). The need for hydrocephalus HT (by either ventriculoperitoneal shunt or endoscopic third ventriculostomy and choroid plexus cauterization (ETV-CPC)) was determined by a pediatric neurosurgeon using clinical and radiographic criteria; a secondary analysis was performed using the MOMS trial criteria for hydrocephalus. The predictive value of each parameter was assessed by ROC-curve and logistic regression analyses.

Fifty affected fetuses were included in the study, of which 32 underwent open hysterotomy and 18 fetoscopic repair. Two cases of neonatal death were excluded from the analysis. The mean gestational ages for the pre-surgery ultrasound, pre-surgery MRI, post-surgery MRI and pre-delivery ultrasound were 21.8 ± 2.1 weeks, 22.0 ±1.8 weeks, 30.4 ±1.6 weeks and 31.0 ±4.9 weeks, respectively. A total of 16 subjects required HT. Area under the curve (AUC) of predictive accuracy for HT showed that HBH grading on post-surgery MRI had the strongest predictive value (0.86; $p<0.01$), outperforming other predictors such as mean ventricular width on pre-surgery US (0.67; $p=0.05$), post-surgery MRI VV (0.73; $p=0.03$), MRI VV growth (0.79; $p=0.01$), change in HBH (0.82; $p<0.01$), and mean ventricular width on pre-delivery US (0.73; $p=0.01$). Other variables such as mean ventricular width on pre-surgery and post-surgery MRI, and ventricular growth assessment by MRI or US, had an AUC<0.7. Optimal cut-offs of the variables with the highest AUCs were evaluated to improve prediction. A combination of ventricular volume growth ≥ 2.02 ml/week and/or HBH of 3 on post-surgery MRI were the optimal cut-offs for the best prediction [OR: 42 (95% CI: 4 - 431), accuracy: 84%]. Logistic regression analyses also showed that persistence of severe HBH 6 weeks after surgery by MRI is one of the best predictors for HT [OR 39 (95% CI: 4 - 369), accuracy: 84%]. There was no significant change in the results when the MOMS trial criteria for hydrocephalus were used as the dependent variable [3].

Sanz-Cortés et al., described and compared placental and amniotic histology in women who underwent a fetoscopic myelomeningocele repair to those who underwent an open-hysterotomy myelomeningocele repair. Also, we intended to compare findings from both prenatal repair groups to age-matched control pregnant patients.

Placental and membrane histopathology from 43 prenatally repaired spina bifida cases (17 fetoscopic and 26 open) and 18 healthy controls were retrospectively assessed. Quantitative assessment of histopathology included apoptosis count, maternal and fetal underperfusion scores. Qualitative assessment included the detection of pigmented macrophages and/or signs of placental/amniotic inflammation. Associations between the duration of surgery or the duration of CO_2 insufflation and quantitative histological parameters were tested.

Fetoscopic surgery cases did not show significant differences in any of the studied parameters when

compared against controls. No differences were detected either when compared to open-repaired cases, except for lower proportion of pigmented laden macrophages in the fetoscopic group (11.8% vs 61.5% p<0.01). No associations between the duration of surgery or the duration of CO2 exposure and any of the quantitative histological parameters were detected.

These preliminary results support the lack of detrimental effects of the use of heated and humidified CO2 gas for uterine insufflation to fetal membranes and placenta [4].

Macedo et al., from São Paulo, selected from the database patients aged 5 years or older for evaluation of urinary and fecal continence. They reviewed all charts and completed a questionnaire to study aspects of urinary and fecal continence.

They identified 14 patients, i.e., 4 (28.6%) males and 10 (71.4%) females. The mean age at MMC surgery was 25.6 gestational weeks. The uro-dynamic class was high-risk in 6 (42.9%), incontinent in 4 (28.6%), hypocontractile in 1 (7.1%), and normal in 3 (21.4%) patients. Three patients had undergone surgery (2 augmentations, i.e., 1 in association with a left colon ACE Macedo-Malone procedure and 1 mini-sling urethroplasty). Twelve patients underwent clean intermittent catheterization (CIC) (85.7%). Only 3 (21.4%) patients had no urinary leakage. Eleven patients (78.6%) used diapers. Eight patients (57.2%) underwent retrograde rectal irrigation and 11 (78.6%) complained of fecal loss. Eleven patients (78.6%) did not report an impact on their self-esteem.

Despite the use of CIC in 85.7% of the cases, the continence rate in MMC patients operated on in utero was low and 78.6% of the patients used diapers. This data can be used to educate parents about future conditions of their 5-year-old children and may stimulate the debate regarding further attempts (surgical or not) to improve fecal and urinary continence [5].

A prospective study of Zarutskie et al., from the Baylor College of Medicine, Texas Children's Hospital, Lucile Packard Children's Hospital Stanford from fetuses diagnosed with open neural tube defect that had in-utero myelomeningocele repair between April 2014 and April 2016. Independent variables were collected from four chronological sets of fetal images: pre-surgery ultrasound, pre-surgery MRI, 6-week post-surgery MRI and pre-delivery ultrasound. The following independent variables were collected from all image sets unless otherwise noted: gestational age, head circumference, mean ventricular width, ventricular volume (VV, MRI only), hindbrain herniation (HBH) score (MRI only), and level of lesion, defined as the upper bony spinal defect (pre-surgery US). Based on these measurements, additional variables were defined and calculated including change in degree of HBH, ventricular width growth (mm/week), and ventricular volume growth (ml/week). The need for hydrocephalus HT (by either ventriculoperitoneal shunt or endoscopic third ventriculostomy and choroid plexus cauterization (ETV-CPC)) was determined by a pediatric neurosurgeon using clinical and radiographic criteria; a secondary analysis was performed using the MOMS trial criteria for hydrocephalus. The predictive value of each parameter was assessed by ROC-curve and logistic regression analyses.

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outperforming other predictors such as mean ventricular width on pre-surgery US (0.67; p=0.05), post-surgery MRI VV (0.73; p=0.03), MRI VV growth (0.79; p=0.01), change in HBH (0.82; p<0.01), and mean ventricular width on pre-delivery US (0.73; p=0.01). Other variables such as 🤣, mean ventricular width on pre-surgery and post-surgery MRI, and ventricular growth assessment by MRI or US, had an AUC<0.7. Optimal cut-offs of the variables with the highest AUCs were evaluated to improve prediction. A combination of ventricular volume growth ≥ 2.02 ml/week and/or HBH of 3 on post-surgery MRI were the optimal cut-offs for the best prediction [OR: 42 (95% CI: 4 - 431), accuracy: 84%]. Logistic regression analyses also showed that persistence of severe HBH 6 weeks after surgery by MRI is one of the best predictors for HT [OR 39 (95% CI: 4 - 369), accuracy: 84%]. There was no significant change in the results when the MOMS trial criteria for hydrocephalus were used as the dependent variable [6].

2018

A single-center retrospective analysis was performed of fetal MRI examinations revealing open spinal dysraphism from 2004 through 2016 with available diagnostic postnatal spinal MR images in conjunction with neurosurgical follow-up findings. Images were reviewed by two board-certified fellowship-trained pediatric neuroradiologists. Relevant clinical data were recorded.

The study of the Department of Radiology and Medical Imaging, Department of Pediatric Neurosurgery, Department of Pediatric Surgery, Department of Biostatistics and Epidemiology, Cincinnati Children's Hospital Medical Center, included 119 fetal MRI examinations of patients with open spinal dysraphism. Myeloceles were found in 29.4% (35/119) of these examinations and myelomeningoceles in the others. All (35/35) myeloceles showed grade 3 (severe) Chiari II malformations. Only 73.8% (62/84) of myelomeningoceles showed grade 3 Chiari II malformation. Clinically significant spinal kyphosis was found in 5.0% (6/119) of fetuses, and all of these fetuses had grade 3 Chiari II malformations. The size of the spinal dysraphic defect had significant positive correlation with lateral ($p < 0.0001$) and third ($p = 0.006$) ventricular size. Mean volume of the myelomeningocele sac was significantly different among Chiari II grades and inversely proportional to Chiari II grade ($p = 0.0009$).

Larger spinal dysraphic defects correlated with increased ventricular size at fetal MRI. All of the fetuses with myelocele or kyphosis had severe Chiari II malformations. Larger myelomeningocele sac size was associated with lower grade of Chiari II malformation, suggesting that myelomeningocele sac formation may be protective against hindbrain herniation. [7].

2016

A total of 145 children underwent MM repair between 2000 and 2004; complete data were available for 133 patients. The probability of 10-year survival was 55%, with 78% of deaths occurring in the first 5 years. Most of the deaths were not directly related to MM; infection and neglect were most commonly described. Lesions at motor level L-2 or above were associated with increased mortality (HR 3.176, 95% CI 1.557-6.476). Compared with repair within 48 hours of birth, surgery at 15-29 days was associated with increased mortality (HR 9.091, 95% CI 1.169-70.698).

Infants in low- and middle-income countries with MM can have long-term survival with basic surgical

intervention. Motor level and age at surgery were significant factors influencing outcome. Education of local health care workers and families to ensure both urgent referral for initial treatment and subsequent access to basic medical care are essential to survival [8].

A retrospective cohort study reports 10-year outcomes and factors affecting survival for infants undergoing MM repair at CURE Children's Hospital of Uganda.

Patients were traced by telephone or home visit. Survival was estimated using the Kaplan-Meier method. Multivariate survival was analyzed using the Cox proportional hazards model, investigating the following variables: sex, age at surgery, weight-for-age at surgery, motor level, and presence and management of hydrocephalus.

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Fifty patients underwent surgery after neurosurgical repair and closure of the placode. A simple guide was generated according to the defect height/width and posterior axillary lines/defect width ratio. These 2 ratios were considered to determine which closure technique (with or without primary repair) should be used for the MMC defect reconstruction.

By using this decision-making guide, 20 of the defects were repaired with various flaps, and those of the remaining 30 patients were repaired with primary closure. In all patients, a successful tension-free 1-stage closure was obtained. Except for 4 patients who had flap reconstruction with partial flap necrosis or minimal flap tip necrosis, healing was uneventful without any complications. There were no additional wound complications during the mean follow-up of 6.8 years (range 5 months to 14 years).

Because of various defect sizes and patient characteristics, no single protocol exists for the reconstruction of MMC defects. The guide suggested here might be effective in deciding which method is suitable for closure of MMC skin defects [10].

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