ABSTRACT

**Background:** Post-surgical evaluation is important for spina bifida patients with low-lying medullary cone including measurement of medullary cone position after being treated. This study aimed to evaluate contributing factors of medullary cone ascending following reconstructive surgery.

**Methods:** This was a descriptive study including low-lying medullary cone spina bifida patients who underwent reconstructive surgery at the Department of Neurosurgery, Cipto Mangunkusumo Hospital, Jakarta, from May 2015 to May 2016. Preoperative and postoperative medullary cone position were evaluated using 1.5 Tesla Magnetic Resonance Imaging (MRI).

**Results:** Eight low-lying medullary cone spina bifida patients who had reconstructive or untethering surgery were included in this study. We found six patients (75%) who had shifting of medullary cone level after surgery. Ascending of medullary cone level to L2 was found in 1 patient (12.5%) of pre-school age group, between 3 – 5 years old. All patients had intradural scar which were shown on post-operative MRI. Primary closure of duramater was performed to all patients. Post-operative MRI assessment at 6 months showed ascending of medullary cone 2 to 3 levels in most patients compared to those who had post-operative MRI assessment in less than 6 months.

**Conclusion:** Age at surgery, surgical technique using primary closure of duramater, and timing of post-operative radiological examination affects shifting of medullary cone position following reconstructive surgery.

**Keywords:** low-lying medullary cone, spina bifida, untethering

INTRODUCTION

Spina bifida occurs in third to fourth week of gestational age. Folic acid deficiency, history of spina bifida in previous child, trisomy 13 and trisomy 18 are the aetiologies of spina bifida. Spina bifida is divided into two types, which are spina bifida aperta and occulta. In this modern era of neurosurgery, spina bifida can also be treated during gestational period, before the delivery.

The common problem for neurosurgeon in treating spina bifida is retethering. Tethered cord syndrome is a collection of symptoms resulted by functional disorder of the spinal cord, which caused by strain of the spinal cord. The strain is occurred due to tethering of the spinal cord in an inelastic structure. The symptoms of tethered cord syndrome can be classified into neurological, orthopaedic, and urological symptoms. Retethering incidence was higher in myelomeningocele compared to lipomyelomeningocele, with incidence rate of 52% and 11%, respectively. The prevalence of retethering was 2.8 – 32%, which majorly cause by post-operative scar. Post-operative scar can be occurred due to direct contact of duramater and neural placode following reconstruction surgery. This leads to lower position of medullary cone than normal. Low-lying medullary cone is one of the tethered cord indicators.

The position of low-lying medullary cones may cause neurological deficits, which possibly manifest during development. Periodical evaluation after reconstructive or untethering treatment in spina bifida patients with low-lying medullary cone is essential as an early detection for tethered cord, because clinical deterioration may potentially occur.

Based on literature review, contributing factors of post-operative medullary cones position shifting were age at surgery, gender, level of spina bifida, spina bifida type, post-operative scar and surgical technique of dural closure. In this study, we aimed to evaluate spina bifida patients with low-lying medullary cone post reconstructive surgery in Department of Neurosurgery, Cipto Mangunkusumo Hospital, Jakarta. In this study, all interventions were performed after delivery.
METHODS
This was a descriptive study including eight spina bifida patients with low-lying medullary cone who underwent repairment reconstructive surgery at Department of Neurosurgery, Cipto Mangunkusumo General Hospital, from May 2015 to May 2016. Data had been descriptively narrated to investigate the factors that may contribute to shifting of medullary cone position following reconstruction surgery in low-lying medullary cone spina bifida, such as age at surgery, gender, level and type of spina bifida, post-operative scar, and dural closure technique.

Pre-operative and post-operative position of medullary cone were analysed using MRI 1.5 Tesla. The position was determined using sagittal section of T2W sequence. The low-lying position of medullary cone was defined as position of medullary cone below L2 level of vertebrae.

Lipomyelomeningocele (LMMC) was defined as a combination of fat, spinal cord, and meninges as cele content during intraoperative observation, while myelomeningocele (MMC) was defined as absence of fat tissue as cele content. We defined scoliosis as lateral spinal curvature more than 10 degrees in pre- or post-operative MRI. Primary dural closure is the closing of dura with only suture without using any tissue graft or synthetic dura. Lastly, post-operative scarring was defined as absence of CSF component between nerve component and dura in axial and sagittal T2W MRI.

RESULTS
Eight patients were included in our study. Preoperative and postoperative MRI examination had been done to assess position of the medullary cone. All of them were patients who were underwent reconstructive surgery for the first time (Table 1).

In this study, samples were dominated by 1 to 3 years of age who underwent the first reconstructive surgery. They were equal between male and female. One patient (12.5%) who was grouped as pre-school aged between 3 – 5 years old had shifting of medullary cone level to L2. Shifting of medullary cone level below L2 were dominated by toddler group between 1 to 3 years old (57.1%), followed by 1 to 11 months of age (28.7%) and pre-school aged group (14.2%). Patients between 1 – 11 months of age showed 1 level post-operative ascending of medullary cone in 2 of 2 patients. In 1 – 3 years of age, 2 levels post-operative ascending of medullary cone was found in one patient of 4 patients. In 3 – 5 years of age, 2 to 3 levels post-operative ascending of medullary cone was found in 2 of 2 patients.

Among 8 patients, seven patients were reported undergoing shifting of medullary cone level below L2, who dominated by four male patients (57.1%) and three female patients (42.9%). Following MRI examination on nine months after surgery, only 1 patient had no ascending of medullary cone. On the other hand, one female patient had ascending of medullary cone level to L2. In seven patients with shifting of medullary cone level below L2, five patients had myelomeningocele (71.4%) and the other two had lipomyelomeningocele (28.57%).

<table>
<thead>
<tr>
<th>No.</th>
<th>Sex</th>
<th>Age at surgery</th>
<th>Medullary cone position</th>
<th>Spina bifida type</th>
<th>Primary dural closure</th>
<th>Post-op scarring</th>
<th>Scoliosis</th>
<th>Clinical findings</th>
<th>MRI post op</th>
<th>Ascent of medullary cone (level)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>15 mo</td>
<td>S3</td>
<td>MMC</td>
<td>Ye s</td>
<td>Ye s</td>
<td>No</td>
<td>No</td>
<td>5 mo</td>
<td>1</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>5 yo</td>
<td>S1</td>
<td>LMMC</td>
<td>Ye s</td>
<td>Ye s</td>
<td>No</td>
<td>No</td>
<td>6 mo</td>
<td>2</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>3 mo</td>
<td>L5</td>
<td>MMC</td>
<td>Ye s</td>
<td>Ye s</td>
<td>No</td>
<td>No</td>
<td>6 mo</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>5 yo</td>
<td>L5</td>
<td>LMMC</td>
<td>Ye s</td>
<td>Ye s</td>
<td>No</td>
<td>No</td>
<td>13 mo</td>
<td>3</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>6 mo</td>
<td>L4</td>
<td>LMMC</td>
<td>Ye s</td>
<td>Ye s</td>
<td>No</td>
<td>Lump</td>
<td>4 mo</td>
<td>1</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>16 mo</td>
<td>S1</td>
<td>MMC</td>
<td>Ye s</td>
<td>Ye s</td>
<td>No</td>
<td>Lump</td>
<td>6 mo</td>
<td>1</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>15 mo</td>
<td>S1</td>
<td>MMC</td>
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<td>Ye s</td>
<td>Yes</td>
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<td>9 mo</td>
<td>0</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>14 mo</td>
<td>L5</td>
<td>MMC</td>
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<td>No</td>
<td>No</td>
<td>Leg weakness, lump, CSF leakage</td>
<td>6 mo</td>
<td>2</td>
</tr>
</tbody>
</table>

M: Male; F: Female; MMC: Myelomeningocele; LMMC: Lipomyelomeningocele; CSF: Cerebrospinal fluid
Level of spina bifida were varied. Among all patients, mostly were at sacral region, marked by 4 patients. Three other patients with shifting medullary cone level below L2 were in lumbosacral region. One patient with ascending medullary cone level to L2 had type lipomyelomeningocele and located in lumbosacral region. In sacral spina bifida, 1 patient had no shifting, 2 patients had ascending one level, and one patient had ascending two levels following reconstructive surgery.

Shifting of medullary cone below L2 level were found in 1 patient with no post-operative scar and six patients (85.7%) with post-operative scar. On the contrary, ascending medullary cone level to L2 was found in 1 patient with post-operative scar. Post-operative MRI examination on 6 months after surgery showed ascending of medullary cone 2 to 3 levels compared to less than 6 months. Medullary cone positions that did not reach L2 were found majorly in patients who did MRI examination more than 6 months after surgery.

**DISCUSSION**

Spina bifida with low-lying medullary cone requires post-operative evaluation to assess retethering of the spinal cord following first reconstructive surgery. We investigated risk factors that contributed to the changes of medullary cone following surgery in spina bifida patients with low-lying medullary cone. They were including age at surgery, gender, region and type of spina bifida, post-operative scar, and surgical technique of dural closure.11

Several studies reported that range of age to perform first reconstructive surgery in low-lying medullary cone spina bifida were between 1 – 4 years old,7,12,13 which accordance with this study. We found that 3 to 5 years of age experienced ascending of medullary cone 2 to 3 levels. We believed that it was affected by body height and elasticity of the spinal cord. However, this hypothesis requires further study.

Data regarding gender in low-lying medullary cone spina bifida following surgery were rarely to find. Samuel et al. found that retethered spinal cord was more common in female with 69 patients (63%).9 Hudgins et al. reported that male and female were equally distributed in their study.11 Similar with our result, we found nearly equal percentage between male and female with post-operative low-lying medullary cone.

According to Oi and Matsumoto, low-lying medullary cone were mostly located in lumbosacral region from L4 to S3 level.14 Patients with medullary cone lies below S1 were prone to experience spinal cord retethering following surgery. We found that most of the patients who have post-operative low-lying medullary cone were having preoperative medullary cone in the sacral region.

Samuel et al. reported that spina bifida type may influence spinal cord retethering, which causing post-operative low lying medullary cone.9 Effect of spina bifida that influence manifestation of spinal cord retethering can be divided into two groups, which are complex and uncomplex etiological group. Complex group is consisted of lipomyelomeningocele, myelomeningocele, and lumbosacral lipoma; while uncomplex group was including fatty filum terminale and split cord malformation.

In this study, we found that underlying aetiology of post-operative low-lying medullary cone were mostly dominated by myelomeningocele, fatty filum terminale, and lipomyelomeningocele, respectively. In accordance to this study, our study revealed that spina bifida type may affect the occurrence of spinal cord retethering.

One of the underlying factors of post-operative retethered is the presence of post-operative scar. Zide et al. described that scar was occurred due to a shallow area of spinal canal, causing a direct contact between neural components and posterior part of duramater. Scarring can be prevented by ensuring that intradural neural component is free and surrounded by cerebrospinal fluid by making a larger space.15,16 In our study, majority of our samples encountered post-operative scar.

Samuel et al. reported that duraplasty technique can prevent retethered cord.9 In accordance with this research, all patients with post-operative low-lying medullary cone were performed primary closure of duramater. There was 1 patient who had elevation of medullary cone to L2 level undergoing primary closure of duramater. Post-operative scar was evaluated with MRI examination, which showed no CSF component between neural components and spinal duramater in MRI sequences.17

According to Bowman et al., evaluation can be started three months following surgery.8 Post-operative MRI examination on more than 6 months had ascending of medullary cone up to 2 to 3 levels as an early assessment compared to patients who conducted the examination less than 6 months.8,18 We propose, for efficacy and effectiveness reason, MRI evaluation should be performed at 6 months, as we can expect significant shifting of medullary cone at that time.

Spina bifida patients with low-lying medullary cone who have undergone reconstructive surgery require further observation following surgery. It needs to be done to assess retethering of spinal cord, wound infection, cerebrospinal fluid leakage and clinical deterioration.19,20 Evaluation can use clinical comparison of pre-operative and post-operative condition with SBNs (Spina Bifida Neurological Scale). Furthermore. It can also be done using pre- and post-operative MRI.14,21,22

**CONCLUSIONS**

Age at surgery, primary closure of duramater, and time of radiological examination after surgery contributes to the changes of medullary cone after surgery. Further research with longer time and larger research subject is required to determine the epidemiology and factors that may affect spina bifida patient with low-lying medullary cone in Indonesia, considering the limitations of this study.

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**CONFLICT OF INTEREST**

Authors declare no conflict of interest regarding this study.

**ETHICAL APPROVAL**

This study has been approved by Research Ethical Committee of Faculty of Medicine Universitas Indonesia with ethical clearance number 689/UN2.F1/ETIK/2016.
AUTHOR CONTRIBUTION

SA is principal author responsible in analyzing results; IR is secondary author responsible in collecting data and analyzing result; HGT, DT, SI, WS, SWN are co-author contributed in discussing study result.

REFERENCES: