RESEARCH ARTICLE

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# Long-term Outcomes of Children with Myelomeningocele and the Quality of Life in Survivors

**ABSTRACT**

**Objective:** Myelomeningocele, a condition that causes chronic health conditions and diminished quality of life, affects not just the children but also their families. Therefore, we comprehensively evaluated the data of 101 children with MMC (myelomeningocele) and aimed to compare the quality of life between children with MMC and their siblings. It is crucial to understand that children with MMS have a diminished quality of life with social and behavioral aspects and health issues, which can be emotionally challenging for them and their families.

**Methods:** In this retrospective study, we collected data from electronic files, ensuring a comprehensive and accurate representation of the participants' medical history. To measure the quality of life, we used the KIDSCREEN 10 instrument, a widely recognized and validated tool in pediatric research.

**Results:** Of the 101 children, 93 were survivors. Comparing the survivors (n=93) with their siblings, survivors had lower HRQoL (health-related quality of life) scores in subdimensions of physical well-being ($p<0.001$), relationships with family ($p<0.001$) and friends ($p<0.001$), school performance and attention ($p<0.001$). On the other hand, the psychological wellness score was higher in survivors than in siblings ($p<0.001$). Most 44 (43.5%) had average mental capacity. The HRQoL score, a measure of the impact of health conditions on a person's overall well-being, was lower in the Chiari type 2 group than in the other survivors ($p=0.035$). Serum and folic acid levels did not correlate with HRQoL measures.

**Conclusions:** This study illuminates the quality of life measures in MMC survivors and the Chiari type 2 group and utilizes new MRI findings, which provide groundbreaking insights into the health conditions and well-being of these populations. These findings are of utmost importance for medical professionals, researchers, and healthcare providers specializing in pediatric care and neurology, as they can significantly impact the treatment and care of these patients.

**Keywords:** Myelomeningocele, Quality Of Life, Chiari Type 2 Malformation, Magnetic Resonance

# Miyelomeningoselli Çocukların Uzun Dönem Sonuçları ve Hayatta Kalanların Yaşam Kalitesi

**ÖZET**

**Amaç:** Kronik sağlık sorunlarına ve yaşam kalitesinin düşmesine neden olan miyelomeningosel, sadece çocukları değil ailelerini de etkilemektedir. Bu nedenle MMC'li (miyelomeningosel) 101 çocuğun verilerini kapsamlı bir şekilde değerlendirdik ve MMC'li çocuklar ile kardeşleri arasındaki yaşam kalitesini karşılaştırmayı amaçladık. MMS'li çocukların sosyal ve davranışsal yönleri ve sağlık sorunları nedeniyle yaşam kalitesinin düştüğünü ve bunun kendileri ve aileleri için duygusal açıdan zorlayıcı olabileceğini anlamak çok önemlidir.

**Yöntem:** Bu retrospektif çalışmada elektronik dosyalardan veri toplayarak katılımcıların tıbbi geçmişinin kapsamlı ve doğru bir şekilde temsil edilmesini sağladık. Yaşam kalitesini ölçmek için pediatrik araştırmalarda yaygın olarak tanınan ve doğrulanan bir araç olan KIDSCREEN 10 cihazını kullandık.

**Bulgular:** Meningomyeloselli 101 çocuktan 93'ü hayatta kaldı. Hayatta kalanlar (n=93) kardeşleriyle karşılaştırıldığında, hayatta kalanların fiziksel iyilik hali ($p<0,001$), aileyle ilişkiler ($p<0,001$) ve arkadaşlarla ilişkiler ($p<0,001$) alt boyutlarında HRQoL (sağlıkla ilişkili yaşam kalitesi) puanları daha düşüktü ($p<0,001$), okul performansı ve dikkat ($p<0,001$). Öte yandan hayatta kalanlarda psikolojik iyilik puanı kardeşlere göre daha yüksekti ($p<0,001$). Çoğu 44 (%43,5) ortalama zihinsel kapasiteye sahipti. Sağlık koşullarının kişinin genel refahı üzerindeki etkisinin bir ölçüsü olan HRQoL puanı, Chiari tip 2 grubunda diğer hayatta kalanlara göre daha düşüktü ($p=0,035$). Serum ve folik asit düzeyleri HRQoL ölçümleriyle korelasyon göstermedi.

**Sonuç:** Bu çalışma yalnızca MMC'den sağ kurtulanlarda ve Chiari tip 2 grubundaki yaşam kalitesi ölçümlerini aydınlatmakla kalmıyor, aynı zamanda bu popülasyonların sağlık koşulları ve refahı hakkında çığır açıcı bilgiler sağlayan yeni MRI bulgularını da kullanıyor. Bu bulgular, bu hastaların tedavi ve bakımını önemli ölçüde etkileyebileceğinden, pediatrik bakım ve nöroloji alanında uzmanlaşmış tıp uzmanları, araştırmacılar ve sağlık hizmeti sağlayıcıları için son derece önemlidir.

**Anahtar Kelimeler:** Meningomyelosel, Chiari Tip 2 Malformasyonu, Manyetik Rezonans.

## INTRODUCTION

Spina bifida is a development and closure defect of the neural tube. Myelomeningocele

(MMC), Alternatively, open spina bifida is the most severe spina bifida associated with malformations (Chiari tip two malformations, hydrocephalus, brainstem, and midbrain abnormalities). Spinal cord injury and brain abnormalities may cause paralysis, sensory dysfunction, and bladder and bowel incontinence. These abnormalities have neurobehavioral consequences (1,2). Other impairments include learning disabilities, memory difficulties, depression, anxiety, and impulsive behaviors. The children with MMC have survival rates increasing up to %70-80. These advances led to the need to research disabilities and their effects on daily life.

Spina bifida, a chronic health condition, significantly affects an individual's quality of life. This term encompasses their position within their cultural and value systems, goals, expectations, and concerns. The impact of spina bifida extends across multiple domains, including psychosocial, physical, and cognitive issues, as well as school attendance and performance. Understanding and addressing these issues is crucial for improving the lives of those with spina bifida (3,4,5).

The study aims to compare these parameters of individuals with or without Chiari malformations, providing a comprehensive understanding of the unique challenges and needs of those with this specific form of spina bifida.

## MATERIAL AND METHODS

Our study, which drew data from the Duzce University School of Medicine database, focused on children with myelomeningocele (n=101). The HRQoL questionnaire was administered to MMC survivors aged between 2 and 18 years, with non-completion of the questionnaire being the only exclusion criterion. Data from the questionnaires were collected via phone calls, and the patients' parents were duly informed about this retrospective study, ensuring the reliability and relevance of our findings.

Our study adhered to the highest ethical standards per the 1964 Helsinki Declaration. The Bioethics Committee of the Medical University of Duzce University approved the study (date: 18.03.2022 number), further attesting to the ethical soundness of our research.

We present all demographics and laboratory and clinic data (sex, age, hydrocephalus, mobility, urinary incontinence, mental status, cranial malformations, serum Vitamin B12, folic acid, and ferritin levels) with MMC. A child and adolescent psychiatrist evaluated the cognitive capacities of the patients as a result of psychometric tests and psychiatric interviews. Some of the participants' (n=58) psychometric examination files were available in the study and were evaluated with a detailed psychiatric examination by a child and adolescent psychiatrist based on a medical board report. When the retrospectively examined files of the cases were examined, it was found that the Denver II developmental screening test was applied to children between the ages of 0-6, and the Kent-EGY intelligence test - Porteus Labyrinths Tasti and Wisc-R Intelligence tests were applied to children and adolescents between the ages of 6-18 to evaluate the cognitive development of some cases. As a result of all these examinations, the cognitive capacity and accompanying psychiatric disorders of the cases were evaluated. The Kent EGY test is applied to evaluate the verbal intelligence skills of individuals based on knowledge and language; there is no time limit, and it is used individually in a single session. We used The Porteus Maze Test and The Denver Developmental Screening Test (DDST) for cognitive capacity. The Porteus Maze Test was used as a nonverbal intelligence test to measure planning and adaptation to new conditions, as well as some competent functions such as making decisions, prescience, impulsivity, and the ability to delay contentment (6). DDST was used to detect developmental retardation in infants and children below 6 years of age. The parameters of this test detect properties such as; gross motor, language, fine motor-adaptive, and personal-social (7).

We also compared the HRQoL between several groups [Chiari Type 2 group vs. non-Chiari Type 2 group) (MMC survivors vs. their siblings)]. The questionnaire included eleven questions, revealing physical, psychological, social status, and school performance. Questions 1-2 are related to general health, and 3 and 4 show emotional levels. Questions 4-9 include social accordance (lifestyle, relationships with family and friends). Finally, questions 10-11 show school attendance, success, and attention (Table 1) (8).

Our data analysis was comprehensive and rigorous. We used IBM SPSS V23 to analyze the data, calculating HLQoL scores by T scores (a mean value of 50 ± a standard deviation of 10). The Kolmogorov-Smirnov test confirmed the normal distribution, and the Mann-Whitney U test was used to compare the groups. Spearman's rho correlation coefficient was employed to analyze the relationship between folate and quality of life. We presented the quantitative data as mean, deviation, median, minimum, and maximum, and categorical data was shown as frequency and percentage. A p-value below 0.05 was considered significant, ensuring the thoroughness and reliability of our findings.

## RESULTS

Out of 101 children with MMC, 93 (92.08%) survived, resulting in a cumulative survival rate of 92%. The average age of the children at the time of the study was 9.2±6.1 years. Fifty-five percent of the children (n=51) were male.

**Table 1.** Kidscreen 10 instrument (6)

| No | Question | Answer | | | | |
|---|---|---|---|---|---|---|
| 1. | Has your child felt fit and well? | Not at all | Slightly | Moderately | Very | Extremely |
| 2. | Has your child felt full of energy? | Never | Seldom | Quite often | Very often | Always |
| 3. | Has your child felt sad? | Never | Seldom | Quite often | Very often | Always |
| 4. | Has your child felt lonely? | Never | Seldom | Quite often | Very often | Always |
| 5. | Has your child enough time for him or herself? | Never | Seldom | Quite often | Very often | Always |
| 6. | Has your child been able to do things in her/his free time? | Never | Seldom | Quite often | Very often | Always |
| 7. | Has your child felt that her/his parents treated him/her fairly? | Never | Seldom | Quite often | Very often | Always |
| 8. | Has your child had fun with his or her friends? | Never | Seldom | Quite often | Very often | Always |
| 9. | Has your child had fun with his or her friends? | Never | Seldom | Quite often | Very often | Always |
| 10. | Has your child got on well at school? | Not at all | Slightly | Moderately | Very | Extremely |
| 11. | Has your child been able to pay attention? | Never | Seldom | Quite often | Very often | Always |

Ninety-two children underwent surgery within one week after birth, and unfortunately, one child passed away on the first day of life. The most common complications found were shunted hydrocephalus (n=71, 70.3%) and Chiari type 2 malformation (n=20, 25.32%). Of the 101 children, 58 (57.8%) were unable to walk, while 21 (20.79%) were able to walk unaided. Approximately 56.4% of the children required clean intermittent catheterization. Most families had 4 (n=27) or 5 (n=20) members.

Cognitive evaluation revealed that 20 (19.8%) had cognitive impairment, and 38 had average cognitive capacity (Table 2). Psychiatric disorders included one (1.8%) adjustment disorder, 6 (10.9%) specific learning disability, and one (1.8%) selective mutism. Also, we report that three (5.5%) had attention deficit hyperactivity disorder, two (3.6%) had an anxiety disorder, two (3.6%) had conduct disorder, and one (1.8%) had a depressive attack.

**Table 2.** Demographics

| | n=101 | % |
|---|---|---|
| Survival | | |
| Live | 93 | 92.08 |
| Dead | 8 | 7.92 |
| Sex | | |
| Male | 51 | 50.50 |
| Female | 50 | 49.50 |
| Hydrocephalus | 71 | 70.30 |
| Chiari type 2 malformation | 20 | 25.32 |
| Mobility | | |
| Unable to walk | 58 | 57.43 |
| Walking with sticks or crutches | 22 | 21.78 |
| Walking unaided | 21 | 20.79 |
| Urinary continence | | |
| Always dry | 12 | 11.8 |
| Always wet | 31 | 30.6 |
| Clean intermittent catheterizations | 57 | 56.4 |
| Unknown | 1 | 0.9 |
| Family size (number of individuals) | | |
| <4 | 6 | 9.67 |
| 4 | 27 | 43.55 |
| 5 | 20 | 32.26 |
| ≥6 | 9 | 14.52 |
| Cognitive function | | |
| Average mental capacity | 38 | 37.6 |
| Mild cognitive impairment | 7 | 6.9 |
| Moderate cognitive impairment | 4 | 4 |
| Severe cognitive impairment | 5 | 5 |
| Mild-moderate cognitive impairment | 1 | 1 |
| Moderate-severe cognitive impairment | 2 | 1.98 |
| Borderline mental retardation | 1 | 1 |
| Unknown | 43 | 42.5 |

The median (minimum-maximum) levels of Vitamin B12, folic acid, and ferritin were approximately 302.2 (47-1500), 26.9 (3.8-518), and 8.5 (1.7-172.6), respectively. The study found no correlation between folate levels and HRQoL (r=-0.122; p=0.519). The most common brain MRI findings were inferior cerebellar vermis and tonsils (n=45, 54.2%), corpus callosum dysgenesis, agenesis (n=37, 44.5%), and colpocephaly (n=27, 32.5%). Spinal MRI findings included tethered cord (n=16, 19%), syringohydromyelia (n=13, 16%), and scoliosis (n=3, 3.75%) (Table 4). Additionally, one case involved situs inversus abdominis, and two involved renal agenesis (Table 3).

**Table 3.** Cranial and spinal malformations

| | n=101 | % |
|---|---|---|
| **Cranial findings:** | | |
| Inferior cerebellar tonsils and vermis | 45 | 54.22 |
| Corpus callosum disgenezis, agenezis | 37 | 44.58 |
| Colphocephaly | 27 | 32.5 |
| Inferior fourth ventricle | 17 | 20.48 |
| Interdigitation of posterior and midline structures | 13 | 15.85 |
| Inferior tentorium | 12 | 14.46 |
| Pecking in the tectum | 8 | 9.64 |
| Brainstem atrophy | 5 | 6.02 |
| Cerebellar atrophy | 2 | 2.41 |
| Vermian hypoplasia | 1 | 1.20 |
| Third ventricle's superiority | 1 | 1.20 |
| Leukomalacia | 2 | 2.41 |
| Tower cerebellum | 1 | 1.20 |
| Septal dysplasia | 2 | 2.41 |
| Absent Septum Pellicidum | 1 | 1.20 |
| The cerebellum migrating to the upper cervical canal | 2 | 2.41 |
| Inferior Medulla oblongata | 1 | 1.20 |
| Evert hippocampus | 1 | 1.08 |
| Superior tentorium | 1 | 1.20 |
| Posterior fossa hypoplasia | 2 | 2.41 |
| **Spinal findings:** | | |
| Tethered cord | 16 | 19.05 |
| Syringohydromyelia | 13 | 16.05 |
| Medullar Spur | 6 | 7.1 |
| Scoliosis | 3 | 3.75 |

Our study, involving a sibling group (n=73) with a mean age of 11.58 and a standard deviation of 7.43, revealed some unexpected findings. Fifty-six percent (n=41) of the siblings were male. When comparing the HRQoL between the siblings and MSS survivors, we found that the siblings had a significantly higher median total score than the survivors (57.61 vs. 43.58, respectively) ($p<0.001$). This difference was also evident in the median scores of physical well-being (58.23 vs. 41.83, $p<0.001$), social functioning (55.86 vs. 43.27, $p<0.001$), friends relations (56.67 vs. 41.31, $p<0.001$), and school performance (58.81 vs. 44.72, $p<0.001$). Even the mean HRQoL score of relations with family was higher in siblings (53.81±5.05) than in survivors (46.81±12.14) ($p<0.001$). However, it was surprising to find that the psychologic status was better in the survivors (median score of 54.8) than in the siblings (median score of 42.68)($p<0.001$)(Table 4). This unexpected finding opens up new avenues for research and warrants further investigation.

The Chiari type 2 group (median of 44.64) had lower HRQoL scores than the non-Chiari type 2 group (median of 50.16) in the MMC survivors group (p=0.035). This finding suggests that individuals with Chiari type 2 malformations may experience more significant impairments in their quality of life than those without this specific condition (Table 5). The psychological evaluation of the health status of some of the patients with myelomeningocele (n=55) was available from the medical records.

**Table 4.** Comparing the HRQoL of MMSS and their siblings

| | MMSS (n=71) | | Siblings (n=73) | | ES | TS* | p |
|---|---|---|---|---|---|---|---|
| | Median (Min-Max) | Mean SD | Median (Min-Max) | Mean SD | | | |
| Total | 43.58 (23.33 – 70.07) | 43.4 ± 9.9 | 57.61 (49.82 – 62.28) | 56.4 ± 4.4 | 1.634 | 619.5 | <**0.001** |
| Physical well-being | 41.83 (25.43 – 58.23) | 42 ± 8.53 | 58.23 (50.03 – 58.23) | 57.78 ± 1.88 | 2.372 | 295.5 | <**0.001** |
| Psychological well-being | 54.8 (42.68 – 91.14) | 56.59 ± 10.31 | 42.68 (42.68 – 66.91) | 43.59 ± 3.3 | 2.202 | 368.5 | <**0.001** |
| Social functioning | 43.27 (22.29 – 55.86) | 44.1 ± 11.56 | 55.86 (47.47 – 55.86) | 55.74 ± 0.98 | 1.207 | 1039.5 | <**0.001** |
| Relationships with family | 55.44 (4.35 – 55.44) | 46.09 ± 12.14 | 55.44 (38.41 – 55.44) | 53.81 ± 5.05 | 0.657 | 1654.5 | <**0.001** |
| Relationships with friends | 41.31 (25.95 – 56.67) | 44.23 ± 11.34 | 56.67 (41.31 – 56.67) | 55.61 ± 2.95 | 1.135 | 1109 | <**0.001** |
| School functioning | 44.72 (37.67 – 65.85) | 46.6 ± 8.71 | 58.81 (37.67 – 65.85) | 53.31 ± 10.12 | 0.695 | 1605.5 | <**0.001** |

ES: Effect Size; T.S.: Test statistics, * Mann Whitney U test, MMSS: Myelomeningocele survivors, HRQoL: Health-related quality of life

**Table 5.** The comparison of HRQoL of the Chiari type 2 malformation group with the non-Chiari type 2 malformation group

| | Non-Chiari type 2 malformation group (n=45) | | Chiari type 2 malformation group (n=16) | | ES | TS* | p |
|---|---|---|---|---|---|---|---|
| | Median (Min-Max) | Mean SD | Median (Min-Max) | Mean SD | | | |
| Total | 50.16 (31.27 – 76.91) | 51.41 ± 9.84 | 44.64 (29.70-61.17) | 45.24 ± 8.30 | 0.560 | 231.5 | **0.035** |

ES: Effect Size; T.S.: Test statistics, * Mann Whitney U test, MMSS: Myelomeningocele survivors, HRQoL: Health-related quality of life

## DISCUSSION

Myelomeningocele causes both physical and mental sequela. The underlying aetiologies are the brain and spinal malformations (mostly hydrocephalus and Chiari type 2 malformations (. Paschereit et al. reported that the histopathological data of the patients with MMC is variable. The malformations were hydrocephalus (71%), Chiari II malformation (36%), heterotopia (34%), cerebellar anomalies (36%), gyrification defects (33%), ependymal denudation (29%), abnormality of corpus callosum (19%). These findings indicate a wide range of brain and spinal cord structural abnormalities, likely contributing to the observed differences in HRQoL scores. Rare anomalies were dysplastic cranial nerves, hypoplasia/aplasia of the mesencephalon, aplasia of olfactory bulbous, atresia of the 4th ventricle, aqueductal stenosis, anomalies of the limbic system, hippocampal anomaly, aplasia of the fornix, nuclear anomalies, dysplastic olives, diminution of pontine nuclei, and the meningeal malformations. Some of these histopathological findings were not present in MRI findings. They concluded that cerebellar malformations and cerebral heterotopias accompany MMC (9). In the literature, cerebellar dysplasia, abnormal gyrus structure in the cerebral hemispheres, colpocephaly, corpus callosum dysgenesis or agenesis, large massa intermedia, small third ventricle, aqueductal anomalies, absent tentorium, displacement of the cerebellum towards the upper cervical canal, pons, herniation of the cerebellar vermis and tonsils into the upper cervical canal are well documented (9, 10). These findings further support the hypothesis that the observed differences in HRQoL scores are due to the underlying structural abnormalities in the brain and spinal cord.

In our study, of 101 patients with MMC, the most common malformations were inferior cerebellar tonsils and vermis (n=45, 54.2%), corpus callosum dysgenesis, agenesis (n=37, 44,5%), colpocephaly (n=27, 32,5%), inferior fourth ventricle (n=17, 20.48%), interdigitation of posterior and midline structures (n=13, %15.85), pecking in the tectum (n=8, 9.64%), brainstem atrophy (n=5, %6.02). More common than the literature, we detected interdigitation of the structures, pecking in the septum, and brainstem atrophy.

Our patients' rare malformations, which were not reported before, were vermian hypoplasia (n=1), third ventricle's superiority (n=1), leukomalacia (n=2), tower cerebellum (n=1), septal dysplasia (n=2), absent septum pallidum (n=1), evert hippocampus (n=1), superior tentorium (n=1), posterior fossa hypoplasia (n=2). In our study, these findings were reported in the literature before: cerebellar atrophy (n=2), cerebellum migrating to the upper cervical canal (n=2), and inferior medulla oblongata (n=1).

Various studies measured the HRQoL in MMC survivors. Buoro et al. reported that myelomeningocele harmed the caregiver's or mother's quality of life, functional capacity, emotional status, and mental health. Bladder catheterization, general care, and financial

limitations are the main problems. They used the World Health Organisation Quality of Life, BREF questionnaire (11). Karmur et al. found that age, level of myelomeningocele, and socioeconomic factors were related to health status outcomes (12). Hydrocephalus Outcome Questionnaire and Health Utility Index scores revealed higher scores of HRQOL associated with lower age, lower anatomical level of myelomeningocele, better family functioning, and higher family income in patients with myelomeningocele and shunted hydrocephalus (12). Previous studies revealed impairment of HRQOL in the domains of health, relationships, esteem and sexuality, bladder, bowel, and bathroom use (13), family and freedom, bladder and bowel esteem, and independence (14).

In another study using Health Utilities Index 3 from Uganda, carer predictions didn't correlate with the child's reported score. Urinary incontinence, essentially family size, male sex, and the presence of hydrocephalus altered the HRQoL in children with MMC. As a component of the WHO QOL index, the Vas score achieved more accurate results than the Health Utilities Index 13) (15). Abanoz et al. compared the HRQoL of children with MMC (n=50) and healthy children (n=50) using the Child Health Questionnaire Parent Form 50. The patients with MMC had significantly lower QOL scores (general health, physical functioning, role/social limitations, emotional/behavioral, mental health, family activities, and family cohesion) than those of the healthy group (14).

Our investigation demonstrated the impairment in MMS survivors' total HRQoL scores and domains (physical well-being, social functioning, friendly relations, family relations, and school performance). Inconsistent with the literature, psychological functioning was significantly better in the MMC survivors than in the siblings. Among the cases whose psychiatric files were accessed, we found that the most common psychiatric diagnoses were mental retardation, specific learning disability and conduct disorder, and attention deficit hyperactivity disorder.

While few studies have examined the impact of Chiari type 2 malformations on quality of life, our study focused on MMC survivors' vision, speech, hearing, dexterity, ambulation, cognition, emotions, and pain. The Chiari type 2 group had worse overall cognition and speech scores, indicating potential cognitive and communication challenges associated with this malformation. We also found a worse total score, including subdimensions of physical well-being, psychological well-being, relationships with family and friends, and school performance in the Chiari Type 2 group, suggesting a broader impact on various aspects of HRQoL.

Our study reveals that HRQoL scores, particularly in the subdimensions of physical well-being, relationships with family and friends, school performance, and attention, are significantly lower in MMS survivors than their siblings. However, MMS survivors surprisingly exhibit higher psychological wellness scores than their siblings. Furthermore, Chiari type 2 malformations significantly exacerbate the already compromised HRQoL scores.

We discovered novel MRI findings in MMS survivors, including vermian hypoplasia, superiority of the third ventricle, leukomalacia, tower cerebellum, septal dysplasia, and absent septum pallidum. These unique findings add a new dimension to our understanding of MMC and its impact on HRQoL. New health policies, home care services and social assistance should be developed to improve the quality of life of meningomyelocele patients.

**REFERENCES**


1. Foss S, Flanders TM, Heuer GG, Schreiber JE. Neurobehavioral outcomes in patients with myelomeningocele. Neurosurg Focus. 2019;47:E6.
2. Bakaniene I, Prasauskiene A, Vaiciene-Magistris N. Health-related quality of life in children with myelomeningocele: a systematic literature review. Child Care Health Dev. 2016;42(5):625-43.
3. Desai VR, Gadgil N, Saad S, Raskin JS, Lam SK. Measures of Health-Related Quality of Life Outcomes in Pediatric Neurosurgery: Literature Review. World Neurosurg. 2019;122:252-65.
4. Sawin KJ, Brei TJ, Houtrow AJ. Quality of life: Guidelines for the care of people with spina bifida. J Pediatr Rehabil Med. 2020;13(4):565-82.
5. Ferro MA, Otto C, Ravens-Sieberer U. Measuring health-related quality of life in young children with physical illness: psychometric properties of the parent-reported KIDSCREEN-27. Qual Life Res. 2022;31(5):1509-20.
6. Porteus SD (1965) Porteus maze tests: fifty years' application. Pacific Books, Oxford [Google Scholar].
7. Frankenburg WK. Denver II – Denver Developmental Screening. Second Version. Nihon Shoni Iji Shuppannsha, Tokyo, 2009.
8. Ravens-Sieberer U, Erhart M, Rajmil L, Herdman M, Auquier P, Bruil J,et al; European KIDSCREEN Group. Reliability, construct and criterion validity of the KIDSCREEN-10 score short measure for children and adolescents' well-being and health-related quality of life. Qual Life Res. 2010;19(10):1487-500.
9. Paschereit F, Schindelmann KH, Hummel M, Schneider J, Stoltenburg-Didinger G, Kaindl AM. Cerebral Abnormalities in Spina Bifida: A Neuropathological Study. Pediatr Dev Pathol. 2022;25(2):107-23.
10. Naidich TP, McLone DG, Fulling KH. The Chiari II malformation: Part IV. The hindbrain deformity. Neuroradiology. 1983;25(4):179-97.

11. Buoro RS, Nogueira MP. QUALITY OF LIFE AND CHALLENGES OF FAMILY MEMBERS OF CHILDREN WITH MENINGOMYELOCELE. Acta Ortop Bras. 2020;28(6):291-5.
12. Karmur BS, Kulkarni AV. Medical and socioeconomic predictors of quality of life in myelomeningocele patients with shunted hydrocephalus. Childs Nerv Syst.2018;34(4):741-7.
13. Szymanski KM, Misseri R, Whittam B, Raposo SM, King SJ, Kaefer M, et al. Quality of Life Assessment in Spina bifida for Adults (QUALAS-A): development and international validation of a novel health-related quality of life instrument. Qual Life Res. 2015;24(10):2355-64.
14. Szymański KM, Misseri R, Whittam B, Casey JT, Yang DY, Raposo SM, et al. Validation of QUALAS-T, a health-related quality-of-life instrument for teenagers with spina bifida. Cent European J Urol. 2017;70(3):306-13.
15. Sims-Williams HJ, Sims-Williams HP, Mbabazi Kabachelor E, Warf BC. Quality of life among children with spina bifida in Uganda. Arch Dis Child. 2017;102(11):1057-61.
16. Şeker Abanoz E, Özmen M, Çalışkan M, Gökçay G, Aydınlı N. Ambulation, lesion level, and health-related quality of life in children with myelomeningocele. Childs Nerv Syst. 2020;36(3):611-6.