Research Article ISSN 2689-1085

Journal of Pediatrics & Neonatology

# Spinal Dysraphism of Lumbosacral Area in Infants: Aspects of Surgical Treatment

**Bodnar Oleh*, Roshka Aurel, Vatamanesku Livii, Randuk Roman, Kyrylyak Sergei and Shevtsiv Ivan**

*Bukovinian State Medical University (Pediatric Surgical Department), Chernivtsy, City, Ukraine.*

***Correspondence:**
Bodnar Oleh, Bukovinian State Medical University, 58002, Ukraine, Chernivtsi, Teatralnaya Square, Tel: +38-050-671-32-83.

**Received:** 08 June 2020; **Accepted:** 27 July 2020

**Citation:** Bodnar Oleh, Roshka Aurel, Vatamanesku Livii, et al. Spinal Dysraphism of Lumbosacral Area in Infants: Aspects of Surgical Treatment. J Pediatr Neonatal. 2020; 2(2): 1-6.

## ABSTRACT

***Background:*** *Spinal dysraphism (SD) is a common birth defect resulting from incomplete closure of the neural tube during the first month of pregnancy. SD at children is difficult and unsolved problem in pediatric neurosurgery. Pathology of the lumbosacral area is 30% - 50% of the total number of SD at children.*

***Objective:*** *To estimate the types, manifestation, complications and to determine the optimal terms and methods of surgical treatment of SD on lumbosacral area in infants.*

***Methods:*** *23 infants with SD were operated. 3 were died after surgery. 20 children were assessed in follow up period (from 1 to 3 years). The children were operated at once after birth to 2 months. Depending on the anatomical variants of SD, the patients were divided into: meningocele (10%); meningoradiculocele (35%); myelomeningocele (30%), myelocystocele (5%) myeloschisis (20%). Findings: 8 children were operated from 1 to 2 days after birth. The best results for surgery were age of 8 days to 2 months (12 children). The late term of surgical operation was due to presence of hernia's coats infection and purulence, which demanded the implementation of preoperative preparation. Cerebrospinal fluid leakage (7 children) has needed to urgent surgery. The disadvantage of all methods of surgical treatment of SD is the danger of damage to neural elements during surgery.*

***Conclusion:*** *During the execution of surgical treatment of SD is necessary to use radiculolysis with precision microsurgical excision of all cicatricial adhesions, cerebrospinal fluid cysts and other intraradicular formations and spinal canal's revision. Dysfunction of the pelvic organs (urine and anal incontinence) in 75% and lower limbs in 40% that observed in children operated on SD in the neonatal period requires further development of methods of their surgical correction at the later age.*

## Keywords

Spinal dysraphism, surgical treatment, infants.

## Introduction

Neural tube defects are the most common malformation of the central nervous system. Spinal dysraphisms are congenital abnormalities of the spinal cord, nerve roots, meninges and vertebra resulting from impaired closure of the mesodermal and ectodermal tissues along the spinal axis during central nervous system development. There are some embryological theories explaining SD occurrence.

- Disorder of primary neurulation

- Nonclosure theory (by von Recklinghausen). It is proposed that neural tube defects represent a primary failure of neural tube closure.

- Overdistension theory (by Morgagni – Gardner). It is proposed that SD arise through overdistension and rupture of a previously closed neural tube (It was confirmed by mouse mutant models).

- Disorder of the midline axial integration during gastrulation (by Dias, Pang and Walker). The failure of the Hensen node to lay down properly a single notochord flanked by a cohesive surrounding sheet of neuroepithelium.

- Failure of disjunction.

During development, the primary neural tube closes at Carnegie stage 12 (30 days, 33 pairs of somites) in the human embryo. Thereafter, the secondary neural tube is formed by a process of differentiation and canalization with the primary neural tube which occurs during stage 13 and ends by stage 20.

The neural tube separates from the overlying ectoderm by a process of disjunction, and the perineural mesenchyme or the somites surround the neural tube and subsequently form the meninges, bone and muscles.

Focal failure of disjunction creates a focal ectodermal-neuroectodermal adhesion and will prohibit the mesenchyme from extending between the neuroectoderm and cutaneous ectoderm and give rise to SD. The neural tube closure and disjunction is a simultaneous process [1].

The SD include spina bifida (SB) and myeloschisis. In the case of the SB, there is the sac containing meninges and cerebrospinal fluid (spina bifida occulta - gap in one or more vertebral arches, but the spinal cord and meninges remain entirely within the vertebral canal; spina bifida aperta).

SB includes different forms regarding to the anatomical concepts.

- **Meningocele.** The sac contains meninges and cerebrospinal fluid, but the spinal cord and spinal root are in their normal position (the skin may or may not be present).
- **Meningoradiculocele.** The sac contains cerebrospinal fluid and neural radices. The nerve roots protrude through the defect in the vertebral arch and are included in the sac.
- **Myelomeningocele.** The meninges as well as the spinal cord (myelon) are found outside of the vertebral arch. The spinal cord lying outside the spinal canal.
- **Myelocystocele.** The dilated (cysto-changed) central canal herniates through a posterior SB defect [2].

Myeloschisis (Rachischisis) is the severest form of the SD. The nerve tissue is here fully bare and a dermal or meningeal covering is absent. With this abnormality, the closure of the neural folds fails to occur. Myeloshisis is defined as an open neural tube defect involving malformation of the vertebral arches, absence of meninges and structural or functional spinal cord abnormalities. Neural placode is located ventrally and is often opposed to the underlying spinal canal. The nerve tissue lies open at the surface without a dermal covering. This leads to the severest disorder of the innervation [3].

Worldwide incidence of the myelomeningocele is 0.2 to 6.4 per 1000 live births with higher rates in less developed countries. Until the early 1950s, many infants born with spinal dysraphism died before reaching adulthood. With better paediatric care, survival rates improved, but little thought was given to longer-term needs. A few paediatricians continued to look after these young people in early adulthood, but most were inevitably subsumed into general clinics. Pathology of the lumbosacral area is 30% - 50% of the total number of SD in children [4].

**The aim of this study is to estimate** the types, manifestation, complications and to determine the optimal terms and methods of surgical treatment of SD on lumbosacral area in infants.

## Materials and Methods

The study was conducted at the paediatric surgical department in Children's hospital in Bukovinian State Medical University (Ukraine, Chernivtsy-city) from 2015 to 2020. There were 23 infants who had SD in the lumbosacral area. 3 children were died after surgery. 1 – spina bifida (L II-L IV) + cerebrospinal fluid leakage (rupture meninges after birth, before surgery) + congenital hydrocephalus, subcompensated type+ lower paraplegia + bladder and bowel incontinence+ cerebral edema+meningitis+pneumonia (death - 12 days after surgery). 2 - spina bifida (L III-L IV) + spina bifida (T I – T IV) + occlusive congenital hydrocephalus, subcompensated type+ convulsion syndrome + respiratory failure + cardiac insufficiency + morfo-functional immaturity of organs and sistems + cerebral edema+ spastic lower paraparesis + bladder and bowel incontinence – (death - 12 days after surgery). 3 - condition after myeloschisis (L I-L IV) repairing – occlusive congenital hydrocephalus, decompensated type; cerebral edema; foramen magnum herniation; acute cardiac insufficiency + flaccid paraparesis + bladder and bowel incontinence (death - 3 months after surgery). Lethality was 13,04%. 20 children were assessed before and after surgical procedures.

### Study Design

Neurological status assessment, optic disk evaluation, ultrasonography (prenatal and postnatal), urological and orthopaedic researches were carried out for all patients. The follow-up was from 1 to 3 years. All children had consent for treatment from their parents in accordance with the principles of the World Medical Association Declaration of Helsinki «Ethical Principles for Medical Research Involving Human Subjects» (amended in October 2013).

Gender, age at surgery, localization and defect size of the SD were recorded in the following Table 1.

| Spinal Dysraphism | Patient | Gender | Age at surgery | Local-ization | Defect size, cm |
|---|---|---|---|---|---|
| Meningocele | 1 | F | 1 month | L | 3×4 |
| | 2 | M | 1,5 month | S | 3,5×5 |
| Meningoradicu-locele | 3 | F | 1 day | L | 7,5×5 |
| | 4 | F | 1 month | L | 3,5×4,5 |
| | 5 | F | 2 days | L | 5,5×7 |
| | 6 | M | 3 months | L | 2,5×5 |
| | 7 | M | 1 day | LS | 6,5×8,5 |
| | 8 | F | 2 months | LS | 3,5×4,5 |
| | 9 | M | 1 month | S | 3×4 |

| | | | | | |
|---|---|---|---|---|---|
| Myelomeningo-cele | 10 | M | 20 days | L | 6x8 |
| | 11 | F | 8 days | L | 7,5x10 |
| | 12 | F | 1 day | LS | 10x11 |
| | 13 | F | 1 month | LS | 5x7 |
| | 14 | M | 1,5 months | S | 4x4 |
| | 15 | M | 2 months | LS | 3x5 |
| Myelocystocele | 16 | F | 1 months | L | 3x5 |
| Myeloschisis | 17 | M | 1 day | L | 6,5x9 |
| | 18 | M | 2 day | LS | 7x10 |
| | 19 | F | 2 day | LS | 5,5x5,5 |
| | 20 | F | 1 day | LS | 8,5x11 |

**Table 1:** The distribution of children according to gender, age at surgery, localization, defect size (n=20).

The children were operated from 1 day to 2 months after birth. Concomitant problems were assessed before and after surgical procedure.

There were some targets of the surgery for SD. Eliminate the foci of pathological impulsation. Restoration of the normal anatomical position and structure of the spinal cord, cauda equina, roots, meninges and surrounding tissue. Inhancement of segmental innervation. Improvement of haemodynamics and CF circulation. Eliminate tethered cord and prevent the tethered spinal cord syndrome in future [5].

There were following stage of the surgery for SD:

- Incision. Cut and hernia sac excision.
- Spinal cord and roots release from surrounding tissue.
- Dura mater is separeted from scar tissue.
- Dural sac is created. It has to be enough space for spinal cord, cauda equina and roots.
- Plastic reconstruction of the dorsal defect of the spinal canal.
- Skin closing

The operation was performed under general anesthesia while the patient was lying in a prone position. Central vein line and bladder catheterization were necessary.

## Description of Procedure

### Incision. Cut and hernia sac excision

The incision was carried caudally and away from the midline along the side to dissect the sac. A circumferential incision was made around the neural placode by saving as much skin as possible. Dura mater was opened by dissecting the skin and the dorsal fascia (Figure 1).

### Spinal cord and roots release from surrounding tissue

Meningoradiculolisis was carried out. If nural roots were thin, atrophic changed it was not possible to detach its from hernia sac (Figure 2).

### Dura mater is separeted from scar tissue

Dura mater was cut laterally and separated from toraco-lumbar fascia. Separation was continued to appearance of the neural roots and epidural veins. Dura mater was mobilized. Dura mater should not have been perforated during the procedure. We did our best to save active functional neural elements. Dissect adhesions (11 cases – adhesions between vertebra and spinal cord) for tethered spinal cord preventing. Spinal cord had to be detached from vertebra (the boned intersections were cut in 6 cases). We tried to check the spinal canal permeability before dura mater closing.

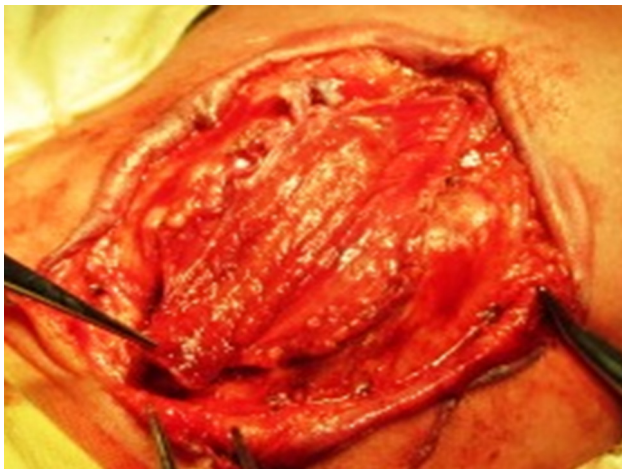

**Figure 1:** Incision. Cut and hernia sac excision.

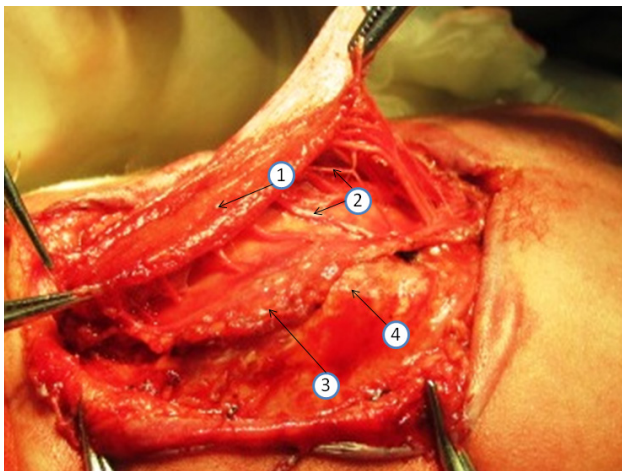


**Figure 2:** Spinal cord and roots release from surrounding tissue (1 - spinal cord; 2 – neural roots; 3 - dura mater; 4 - vertabral).

### Dural sac is created. It has to be enough space for spinal cord, cauda equina and roots

Dura mater was sutured by uninterrupted suture longitudinally (PDS 6\0). The terminal cistern has to be spacious and watertight ! Sometime (5 children), the dura mater duplication was performed (Figure 3).

### Plastic reconstruction of the dorsal defect of the spinal canal

Dura tube were closed by fascio-muscular flaps from toraco-lumbar fascia surrounding muscles (spine muscles or gluteus maximus) (Vicril 3\0). Flaps were rotated on $180^0$ by Bayer. Closing of the dura tube should be without tension. If kyphosis was present – the vertebral osteotomy was done (7 cases).

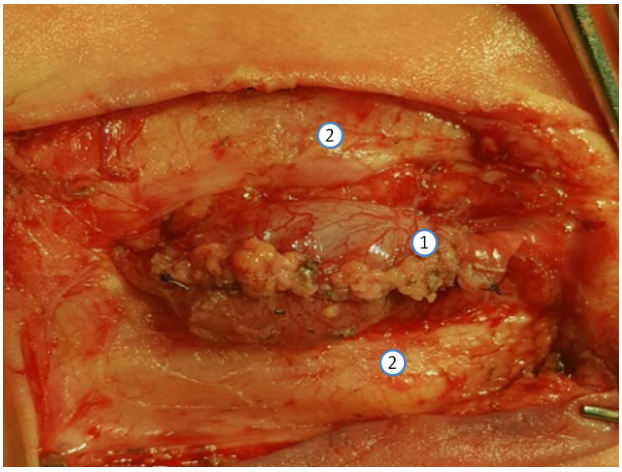


**Figure 3:** Dura mater stitching (1- dura mater; 2 - vertebrae).

From our point of view, Bayer's plasty has some disadvantages. Dissection, separation and rotation of the fascio-muscular flaps led to violation of trophical processes in its, vertebral column deformation, covering only bony defect. If the flaps have necrosis → inflammation → protection fragility → meningitis. Inflammation and scarring → spinal cord and roots include into its → neurological disorders will be strengthening. There is a threat of neurological elemens injury during the surgery.

## Skin closing

Regarding to the skin closing, children were split into two groups. I group. There were 9 (45%) patients who had defect size (DS) < 5 cm. Conventional skin plasty was performed for them. Flaps were mobilized laterally to the midline of the abdomen and stitching above the place of SD plasty (It has to be without tension, but it is not always possible).

II group. There were 11 (55%) kids who had defect size (DS) > 5 cm. Defect repair by bilateral fasciocutaneous flaps (with perforator vessels) rotation by Mustafa Kŭrşat Evrenos, Haldun Onuralp Kamburoglu (Turkey, 2017) was done for them [6], (Figure 4, Figure 5).

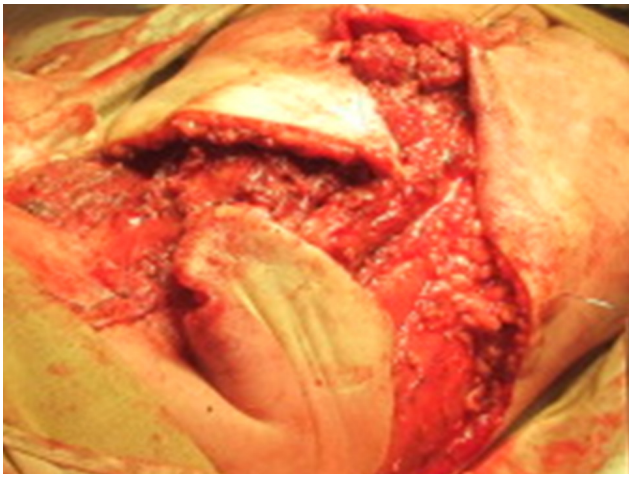

**Figure 4:** Defect repair by bilateral fasciocutaneous flaps (with perforator vessels) rotation.

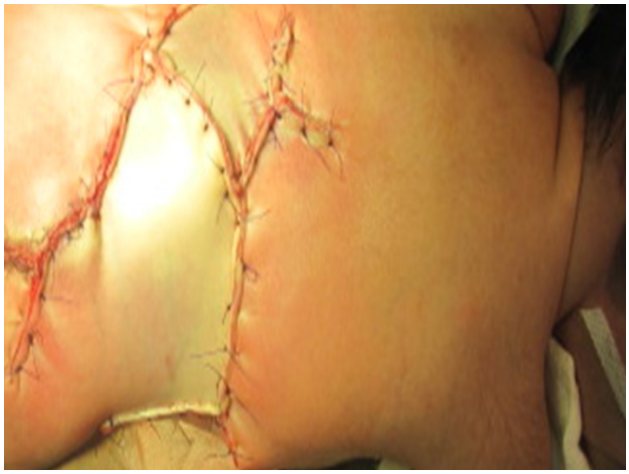

**Figure 5:** Skin closing.

Histological studies (n=20) of the skin near SD defects have identified a lot of vessels and a little of nerves (Figure 6). This demonstrated the possibilities of implementation bilateral fasciocutaneous flaps for this category of patients.

The aim of surgical procedure was to cover the defect with tension-free soft tissues in order to create durable, reliable soft tissue closure while maintaining robust blood supply.

Incision 1 was performed from upper point of wound oblique, laterally in the up. There was toraco-lumbar fascia in the deep of wound.

Incision 2 was performed from lower point of wound oblique, laterally down.

Superiorly dorsal intercostal perforator vessels and inferiorly lumbar perforators were isolated and protected, especially at the base of the flaps.

We used superior flap and inferior flap according to Jeong Tae Kim conception, South Korea, 2005 [7] (Table 2).

| Superior flap (vessels) | Inferior flap (vessels) |
|---|---|
| Latissimus dorsi perforator flap | Latissimus dorsi perforator flap |
| Thoracodorsal perforator flap | Thoracodorsal perforator flap |
| Rectus abdominis lateralis perforator flap | Rectus abdominis lateralis perforator flap |
| | Superior/inferior gluteus maximus perforator flap |

**Table 2:** Defect repair by bilateral fasciocutaneous flaps (with perforator vessels) rotation according to Jeong Tae Kim conception, South Korea, 2005.

Superior flap and inferior flap were rotate (turnover) in opposite positions (up to down, down to up). Approximations of the flaps to the midline were done. The mobilization of flaps should be equal for wound tension. Interrupted sutures (PDS 5\0, PDS 4\0) were used for closure (Figure 6).

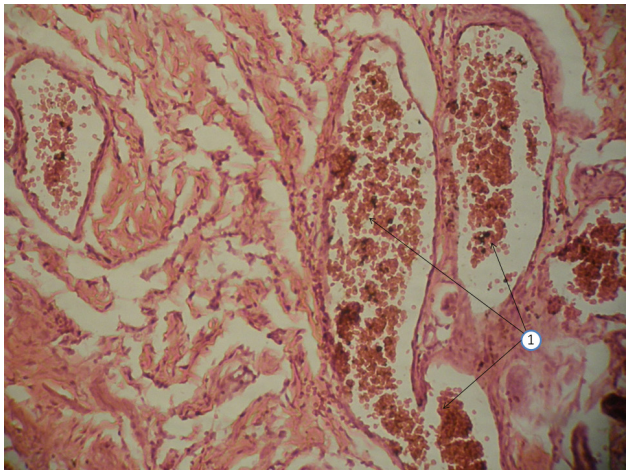


**Figure 6:** Skin near of the spina bifida defect (1 - vessels) (histological research, hematoxylin-eosin staining).

Tension-free soft tissues, protection of the perforator vessels, adequacy of the flaps for wound closing were circumstances for this kind of surgical procedure.

There were indications for the urgent surgeries in infants with SD:

- rupture of the hernia sac + cerebrospinal fluid leakage (CFL) (within first hours but not late than 48 hours);
- membrana of the hernia sac making dramatic thin, or ulceration;
- increase in the size of the hernia sac (increasing number of cerebrospinal fluid);
- myeloschisis.

In other cases, we tried to operate children until 2 months after birth (Table 3).

| Spinal Dysraphism | Time of the surgery after birth | | | | | Total, (n=20) |
|---|---|---|---|---|---|---|
| | 1-2 days | 8 days | 20 days | 21 day - 1 month | 1-2 months | |
| Meningocele, (n=2) | | | | 2 | | 2 |
| Meningoradiculocele, (n=7) | 3 (CFL) | | | 2+1 (CFL) | | 7 |
| Myelomeningocele, (n=6) | 1 (CFL) | 1 (CFL) | 1 (CFL) | 3 | | 6 |
| Myelocystocele, (n=1) | | | | 1 | | 1 |
| Myeloschisis, (n=4) | | | | | | 4 |
| Total, (n=20) | 8 (40%) | 1 (5%) | 1 (5%) | 9 (45%) | 1 (5%) | 20 |

**Table 3:** The distribution of children according to time of the surgery (n=20). CFL: Cerebrospinal Fluid Leakage.

The children with SD had different concomitant problems. They were outcomes of the neural tube defects (Table 4).

Followed up (additional) surgeries were performed for some of this patients. Shunt placement in the cases of hydrocephalus (16 (80%)). Hernioplasty for umbilical or inguinal hernia (4 (20%)). Orthopaedic surgeries (11 (55%)) for congenital clubfoot (4), arthrogryposis (4), hip dysplasia (3). Colon resection and colofixation (1 (5%)) were carried out for prolapse of the rectum, ulcers proctosigmoiditis, dolichocolon.

| Concomitant problems | MC, (n=2) | MRC, (n=7) | MMC, (n=6) | MCC, (n=1) | MSH, (n=4) | Total, (n=20) |
|---|---|---|---|---|---|---|
| Hydrocephalus | 1 | 6 | 5 | | 4 | 16 (80%) |
| Cerebral edema | | 1 | | | | 1 (5%) |
| Meningitis | | 2 | | | | 2 (10%) |
| | | 4 | 3 | | | 7 (35%) |
| Lower paraplegia | | 2 | 2 | | 3 | 7 (35%) |
| Lower paraparesis | | 2 | 2 | 1 | 1 | 6 (30%) |
| Bladder and bowel incontinence | 1 | 6 | 6 | 1 | 4 | 18 (90%) |
| Arnold-Chiari malformation | | 1 | 2 | | 1 | 4 (20%) |
| Kyphosis | | 3 | 3 | | 4 | 10 (50%) |
| Congenital clubfoot | | 3 | 1 | | 1 | 5 (25%) |
| Arthrogryposis | | 1 | 2 | | 2 | 5 (25%) |
| Hip dysplasia | | 3 | 2 | | | 5 (25%) |
| Prolapse of the rectum, ulcers proctosigmoiditis, dolichocolon | | | | | 1 | 1 (5%) |
| Inguinal hernia | | 1 | 1 | | | 2 (10%) |
| Umbilical hernia | 1 | | | 1 | | 2 (10%) |
| Kidney's agenesis | | 1 | 1 | | | 2 (10%) |
| Fused kidney | | 1 | 1 | 1 | | 3 (15%) |
| Pneumonia | | | 1 | | | 1 (5%) |

**Table 4:** Concomitant problems in children operated on SD (n=20).
MC: Meningocele; MRC: Meningoradiculocele; MMC: Myelomeningocele; MCC: Myelocystocele; MSH: Myeloschisis; CFL: Cerebrospinal fluid leakage.

There were the complications after surgery regarding to the groups distribution. They are in the Table 5.

| Complications | Groups of patients (n=20) | |
|---|---|---|
| | I (DS<5cm) (n=9) | II (DS>5cm) (n=11) |
| Wound dehiscence (Secondary healing) | 3 | 1 |
| CFL from incision | 1 | - |
| Subcutaneous CFL collection | - | 1 (Shunt placement) |
| Total | 4 | 2 |

**Table 5:** Complications after surgery (n=20).
CFL: Cerebrospinal Fluid Leakage; DS: Defect Size.

Wound dehiscence in the case of I groups of patients in 33,33% explained by the tension of tissue, lack of the blood supply in the part of the stiches, violation of the microcirculation and joining inflammation. These compromised factors considerably decreased in the case of defect repair by bilateral fasciocutaneous flaps (with perforator vessels) rotation. Only 9,09% were the same complications. Cerebrospinal fluid leakage in the I group were

secondary. It was attributable to insufficient protection of the created dural tube (11,11% of the children).

Monitoring of children during 3 years post-operative period showed improvement regarding to lower paraplegia and lower paraparesis. But, only in 3 from 18 children were improvement bladder and bowel function. Shunt placement was done for 16 patients (Table 6).

| SD+Concomitant problems | Before surgery, (total, n=20) | Improvements after surgery (total, n=20) | Without improvements after surgery (total, n=20) |
|---|---|---|---|
| Hydrocephalus | 16 (80%) | 16 (80%) | - |
| Lower paraparesis | 7 (35%) | 3 (15%) | 4 (20%) |
| Lower paraparesis | 6 (30%) | 2 (10%) | 4 (20%) |
| Bladder and bowel incontinence | 18 (90%) | 3 (15%) | 15 (75%) |

**Table 6:** Post-operative period improvements (n=20).

## Conclusions

- Precision microsurgical technique using with careful relevance to neural roots, blood vessels and surrounding structures provides decrease post-operative clinical manifestation of SD in children.
- SD defect repair by bilateral fasciocutaneous flaps (with perforator vessels) rotation provides improvement of quality of surgical treatment and decrease quantity of post-operative complications.
- The term of surgery for SD in neonates depends on anatomical types of the disease and presence of the complications.
- Concomitant problems, quantity and severity of clinical manifestation of SD in children decrease in the cases of meningocele and increase in the cases of myeloschisis.
- Further researches should be aimed at finding surgical repair of the bladder and bowel incontinence after SD surgical correction.


## Acknowledgment

**Funding:** No funding was requested or given for this registry study. The patients were not paid for participating in the trial, but received free treatments. The information of the manuscript was presented at the Conference Series and the Editors of Neonatal and Pediatric Medicine: Open Access, Pediatrics & Therapeutics and Journal of Pregnancy and Child Health, "13th World Pediatric Congress", December 05-06, 2019 in Rome, Italy.


## References


1. Moududul Haque, Kenji Ohata, Toshihiro Takami, et al. Development of Lumbosacral Spina bifida: Three-Dimensional Computer Graphic Study of Human Embryos at Carnegie Stage Twelve. Pediatric Neurosurgery. 2001; 35: 247-252.
2. Torteri-Donati P, Rossi A, Cama A. Spinal dysraphism: a riview of neuroradiological features with embryological correlations and proposal for a new classification. Neuroradiology. 2000; 42: 471-491.
3. Mehmet Dumlu Aydin. Repair of Myeloschisis Defect with Tutoplast. Turk. J. Med. Sci. 2003; 33: 183-186.
4. Pippa Oakeshott, Alison Poulton, Gillian M. Hunt, et al. Walking and living independently with spina bifida: a 50-year prospective cohort study. Developmental Medicine & Child Neurology. 2019.
5. Gerald F. Tuite, Dominic N.P. Thompson, Paul F. Austin, et al. Evaluation and management of tethered cord syndrome in occult spinal dysraphism.
6. Recommendations from the international children's continence society. Neuroulogy and Urodynamics. 2017; 1-14.
7. Mustafa Kürşat Evrenos, Haldun Onuralp Kamburoğlu, Mehmet Seçer, et al. Clinical Outcomes of Large Meningomyelocele Defect Repair by Bilateral Fasciocutaneous Rotation and Advancement Flaps with Perforators. Turk. J. Plast. Surg. 2017; 25: 113-119.
8. Jeong Tae Kim. New nomenclature concept of perforator flap. British Journal of Plastic Surgery. 2005; 58: 431-440.