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Paper Accepted* **ISSN Online 2406-0895**

Case Report / Приказ болесника

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Anesthesia in infant with spinal muscular atrophy type 1 for ventriculoperitoneal shunt placement

Анестезија код одојчета са спиналном мишићном атрофијом тип 1 за пласирање вентрикуло-перитонеалног шанта

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Received: January 12, 2025
Revised: June 20, 2025
Accepted: July 1, 2025
Online First: July 2, 2025
DOI: https://doi.org/10.2298/SARH250112051N

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SUMMARY

Introduction Spinal muscular atrophy (SMA) is a rare inherited disease in the pediatric population. The most important risk factor associated with anesthesia is increased sensitivity to opioids and neuromuscular blockers as well as problems with airway management and breathing. We present a case report of an eight-month-old female infant undergoing ventriculoperitoneal shunt placement.

Case Outline The infant underwent ventriculoperitoneal shunt placement for hydrocephalus following nusinersen intrathecal injection. The premedication with benzodiazepines are omitted. Except for SMA, the child was considered without comorbidity. The general endotracheal anesthesia was managed with inhalational sevoflurane. The opioid analgesics as well as rocuronium for neuromuscular blockade in a single dose were administered to the child. After the surgery, the child was awake in the operating theater.

Conclusion General endotracheal anesthesia in infants with spinal muscular atrophy can be safely performed. Intravenous anesthesia as well as inhalational anesthesia with sevoflurane are acceptable possibilities.

Keywords: spinal muscular atrophy; anesthesia; inhalational; ventriculoperitoneal shunt

САЖЕТАК

Увод Спинална мишићна атрофија (СМА) је наследна болест која се ретко дијагностикује код деце. Најважнији фактор ризика који је повезан с анестезијом код ове деце је повећана осетљивост на опиоидне аналгетике и неуромишићне релаксанте као и проблеми с обезбеђењем дисајног пута и дисањем. Приказујемо случај осмомесечног женског одојчета код кога је пласиран вентрикулоперитонеални шант.

Приказ болесника Одојчету је планирано пласирање вентрикуло-перитонеалног шанта због хидроцефалуса насталог као последица интратекалне ињекције нусинерсена. Уобичајена премедикација бензодиазепинима није дата детету. Изузев СМА, дете није имало других коморбидитета. Општа ендотрахеална анестезија је одржавана инхалационим севофлураном. Опиодини аналгетици, као и рокуронијум у једној дози су дати детету. По завршетку операције, дете је пробуђено на операционом столу.

Закључак Општа ендотрахеална анестезија код одојчади са спиналном мишичном атрофијом се може извести безбедно. Интравенска анестезија као и инхалациона анестезија са севофлураном су прихватљиве могућности погодне за извођење.

Кључне речи: спинална мишићна атрофија; анестезија; инхалациони; вентрикуло-перитонеални шант

INTRODUCTION

Spinal muscular atrophy (SMA) type 1 is a rare inherited disease in a pediatric population [1]. It is characterized by muscular hypotonia and delayed motor development [2]. General endotracheal anesthesia is challenging in these children because of potential problems with the airway and breathing as well as increased sensitivity to opioids and neuromuscular blockers [3]. We present a case report of an eight-month-old female infant who underwent ventriculoperitoneal shunt placement.

CASE REPORT

An 8-month-old female infant with the diagnosis of SMA type 1 (Werding- Hofman) was admitted to Neurosurgery Clinic, University Clinical Center of Serbia in Belgrade for treatment of obstructive hydrocephalus as a consequence of intrathecal administration of nusinersen. We obtained informed consent from the mother to publish the infant case, with total protection of the patient's data.

This is the fourth child from the fourth pregnancy which was appropriately controlled and uncomplicated. The child was born in the 39th gestational week, by vaginal delivery. At birth, the child had a body weight of 3820 g and a body length of 54 cm, with a head diameter of 37cm and an Apgar score of 9/10. During pregnancy, the mother has noticed that “the baby was not so active”. After birth, except for reduced movements, the parents did not notice any suspicious in the infant.

On regular pediatric control, the pediatrician noticed the presence of hypotonia, so the infant was sent to a pediatric neurologist for examination. On the 43rd day of life, the child was admitted to the pediatric neurology department for evaluation of severe hypotonia. Because of suspicion of SMA type 1, molecular and genetic consultation is indicated to determine the state of mutation in the SMN1 gene. Genetic evaluation revealed that there is homozygous deletion of SMN1 genes and two copies of SMN2 genes. The diagnosis of spinal muscular atrophy type 1 was done. After the confirmed diagnosis of SMA type 1, the indication for nusinersen administration according to protocols was made.

On the 48th day of life, the first dose of nusinersen, 12 mg, was applied intrathecally, and the intervention was done without complications. The first of three doses was administered at 14-day intervals, and the fourth dose was administered 30 days after the third dose. After the fourth dose, the parents noticed an enlarged head circumference, so the magnetic resonance imaging

(MRI) of the cranium was done. It showed the presence of hydrocephalus. The neurosurgeon indicated ventriculoperitoneal shunt placement. The pediatric neurologist indicated interruption of nusinersen intrathecal therapy, but risdiplam in one daily dose of 0.2 mg/kg per os was initiated. The infant was transferred to a tertiary referent neurosurgical center.

On admission, the infant had a body mass weight of 9 kg and a head circumference of 40 cm with fontanela major 3 × 3 cm in the level of cranium bones. The infant was awake, and interested in the environment, with generalized weakness, preserved swallowing, and cough. She was unable to sit independently and to crawl. The pediatric neurologist indicated a continuation of risdiplam therapy, as well as on the day of surgery. She was examined by a pediatrician, and there were no contraindications for surgery. There was no recommendations for preoperative medical respiratory support, the child was ASA score 3. In-room air, the oxygen saturation of hemoglobin was 100% (Figure 1).

Premedication was intentionally omitted due to SMA. Standard monitoring of electrocardiography, non-invasive blood pressure, and pulse oximetry was done. Arterial blood pressure was 125/84 mmHg and heart rate 130/min. The infant was introduced into general endotracheal with O_2/air mixture and sevoflurane at 8 vol% which decreased to 3 vol% till the intubation. After the infant became unresponsive, we started with manual bag ventilation. The nurse inserted and fixed an intravenous cannula. Before intubation, fentanyl 25 mcg and rocuronium 5 mg intravenously were administered and were not repeated. The orotracheal intubation was done with endotracheal tube number 4, with a non-inflated cuff, fixed on 11 cm. The position of the tue was confirmed with chest auscultation and capnography. The throat pack was done. After intubation, mechanical ventilation was initiated (pressure-controlled ventilation with inspiratory pressure of 15cm H2O and end-expiratory pressure of 5 cm H2O, respiratory rate 25/min, FiO_2 50%). End-tidal CO2 was maintained between 37 and 41 mmHg.

Anesthesia was maintained with sevoflurane between 2 and 3 vol% and a total O_2/air flow of 3 L/min. Antibiotic prophylaxis was done with ceftriaxone 500 mg in 100 mL of normal saline infusion.

As usual, the ventriculoperitoneal shunt was placed on the right side: the skin and soft tissue were cut on the parietooccipital part of the scull on the right side (Figure 2). The burr hole was formed. After the cut of the dura mater, the cranial part of the catheter is put into the right lateral ventricle. Clear cerebrospinal fluid occurred in the stream. The catheter is then connected to the pump for medium pressure. The distal part of the system is placed into the peritoneum, paraumbilically, on the right side. The system is functional. Then the suture of the layers is done.

Sevoflurane was stopped and the oxygen was turned to 80% on emergence. The infant was manually ventilated. Neuromuscular blockade was reversed with neostigmine 0.5 mg and atropine 0.2 mg intravenously. After a few minutes, she became fully awake and was crying loudly. Extubation was done in inspirium, without endotracheal suction. After extubation, SpO_2 was 99–100% on oxygen flow 4l/min and 97–98% on room air. The infant was transferred to the intensive care unit for further monitoring and oxygenation. Antibiotic therapy with ceftriaxone once a day was continued. Analgesia was provided with acetaminophen 150 mg (15 mL) intravenously every six hours. Intravenous infusion in equal parts of 10% dextrose solution and normal saline, 40 mL/h was administered.

The next day, she was transferred to the neurosurgical ward in good condition, awake, eupneic, acyanotic, and afebrile. The therapy with risdiplam was continued postoperatively. On the second day after surgery, the infant developed inspiratory stridor. After pediatrician examination, the next therapy is administered: methylprednisolone 1mg/kg intravenously every 12 hours and budesonide inhalations, 0.5 mg diluted with 2 mL of normal saline, every 12

hours. In laboratory findings, C-reactive protein was 11.5 mg/L, and white blood cells were 17.2 x10^9/mL. Antibiotic therapy with ceftriaxone was continued. On the fourth day after surgery, the stridor was in resolution, white blood cells were 17.1 x10^9/mL, and C-reactive protein was 4.8 mg/mL. On the fifth postoperative day, the infant was discharged from the hospital.

Ethics: We obtained informed consent from the mother to publish the infant case, with total protection of the patient's data.

DISCUSSION

Spinal muscular atrophy type 1 is a rare inherited recessive genetic condition diagnosed in the first months of life. A homozygous deletion of SMN1 exon 7 is the most common cause of SMA type 1 and causes the clinical picture of SMA [3]. This disease if untreated, leads to progressive generalized weakness. Affected children usually have hypotonia, difficulty swallowing, and cough, which can result in aspiration pneumonia and respiratory failure [2].

Anesthetic care for infants with SMA type 1 presents significant challenges, and literature on optimal strategies remains limited [1–6, 8, 9]. Anesthesia in this population can be safely performed when individualized care is guided by comprehensive risk assessment tools, such as preoperative pulmonary function evaluation (when feasible), swallowing assessments, and multidisciplinary planning. Detailed history regarding respiratory infections, feeding difficulties, and ventilatory support (e.g., use of non-invasive ventilation at home) is essential to stratify perioperative risk.

There is no universally accepted anesthetic regimen for SMA type 1; however, certain principles are broadly recommended. The use of depolarizing neuromuscular blockers such as

succinylcholine is contraindicated due to the risk of hyperkalemia, rhabdomyolysis, and cardiac arrest, which is related to the upregulation of extrajunctional acetylcholine receptors [3, 5]. Non-depolarizing neuromuscular blockers, such as rocuronium or cisatracurium, can be used cautiously at reduced doses, ideally guided by neuromuscular monitoring (e.g., train-of-four). However, the reliability of such monitoring in SMA patients is debated due to altered neuromuscular transmission [5]. Some authors advocate avoiding muscle relaxants entirely and relying on deep inhalational or intravenous anesthesia [5, 6].

Regarding analgesia, opioid sensitivity is a concern due to impaired clearance and weakened respiratory drive. Short-acting opioids like remifentanil are preferred intraoperatively because of their rapid metabolism and minimal residual effect [2, 8]. Postoperative pain control can then be managed with multimodal analgesia strategies to minimize opioid requirements.

Both intravenous (e.g., propofol) and inhalational (e.g., sevoflurane) anesthetic agents have been used effectively in this population. There is no clear evidence favoring one method over the other in terms of safety or outcomes [2, 5, 9]. The choice often depends on institutional protocols and the anticipated need for postoperative ventilation.

Respiratory management is a central concern in SMA type 1, both pre- and postoperatively. Preoperative preparation should include assessment of airway patency, history of aspiration, and baseline respiratory support. Techniques such as chest physiotherapy, suctioning, and optimization of non-invasive ventilation (NIV) are beneficial. Some institutions advocate preemptive admission to intensive care and early involvement of respiratory therapists.

Intraoperatively, gentle ventilation strategies should be employed to avoid barotrauma, and extubation decisions must consider the child's ability to maintain airway patency and effective ventilation. Postoperative respiratory complications, such as aspiration, atelectasis, or stridor (as seen in our case), may require escalation to non-invasive or invasive mechanical ventilation.

In our case, the development of inspiratory stridor necessitated the use of systemic and inhalational corticosteroids, as well as continued antibiotic therapy.

Importantly, postoperative care should involve close collaboration with pediatricians, pulmonologists, and anesthesiologists, emphasizing the need for a multidisciplinary approach. These children should ideally be treated in tertiary care centers equipped to manage complex airway and neuromuscular issues.

General endotracheal anesthesia can be safely administered to infants with SMA, provided appropriate precautions are taken. Both intravenous and inhalational anesthesia using agents such as sevoflurane are acceptable options. Anesthesia management should include the use of reduced doses of non-depolarizing neuromuscular blockers and short-acting opioids, tailored to the child's neuromuscular status and respiratory function.

Conflict of interest: None declared.

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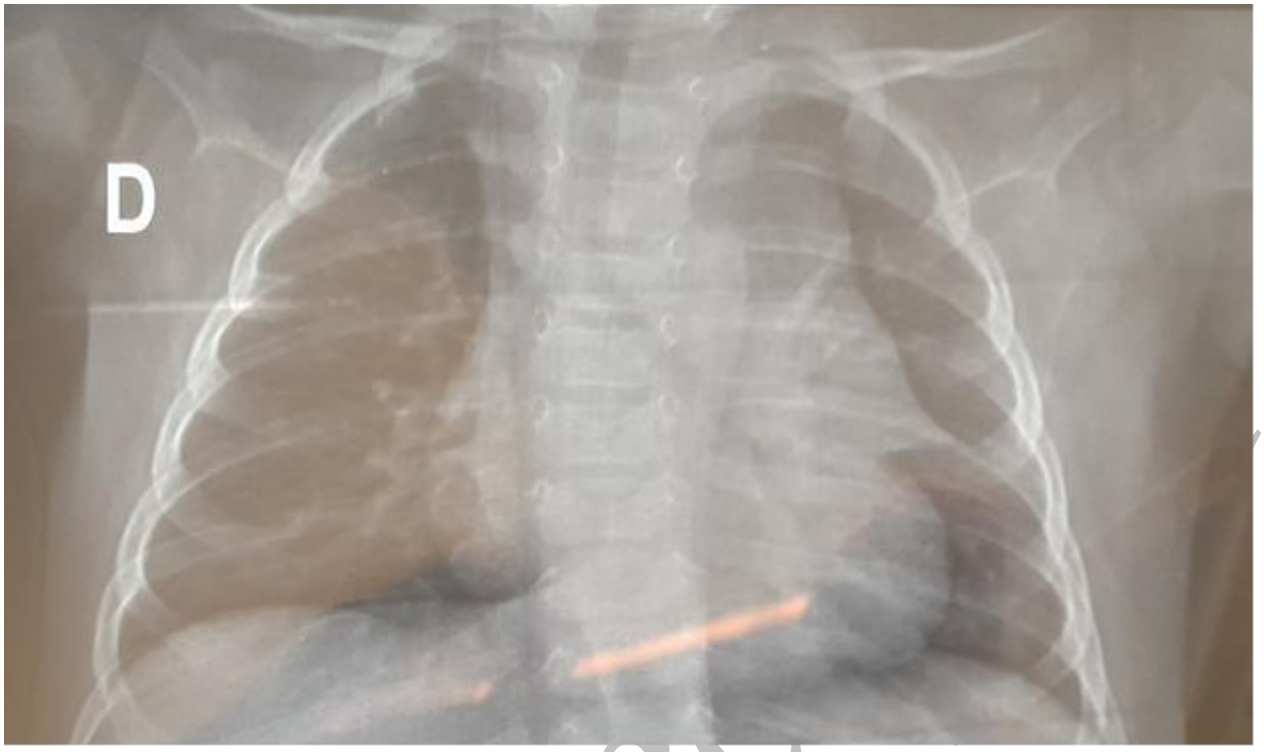

Figure 1. Preoperative chest X-ray in the infant with spinal muscular atrophy type 1

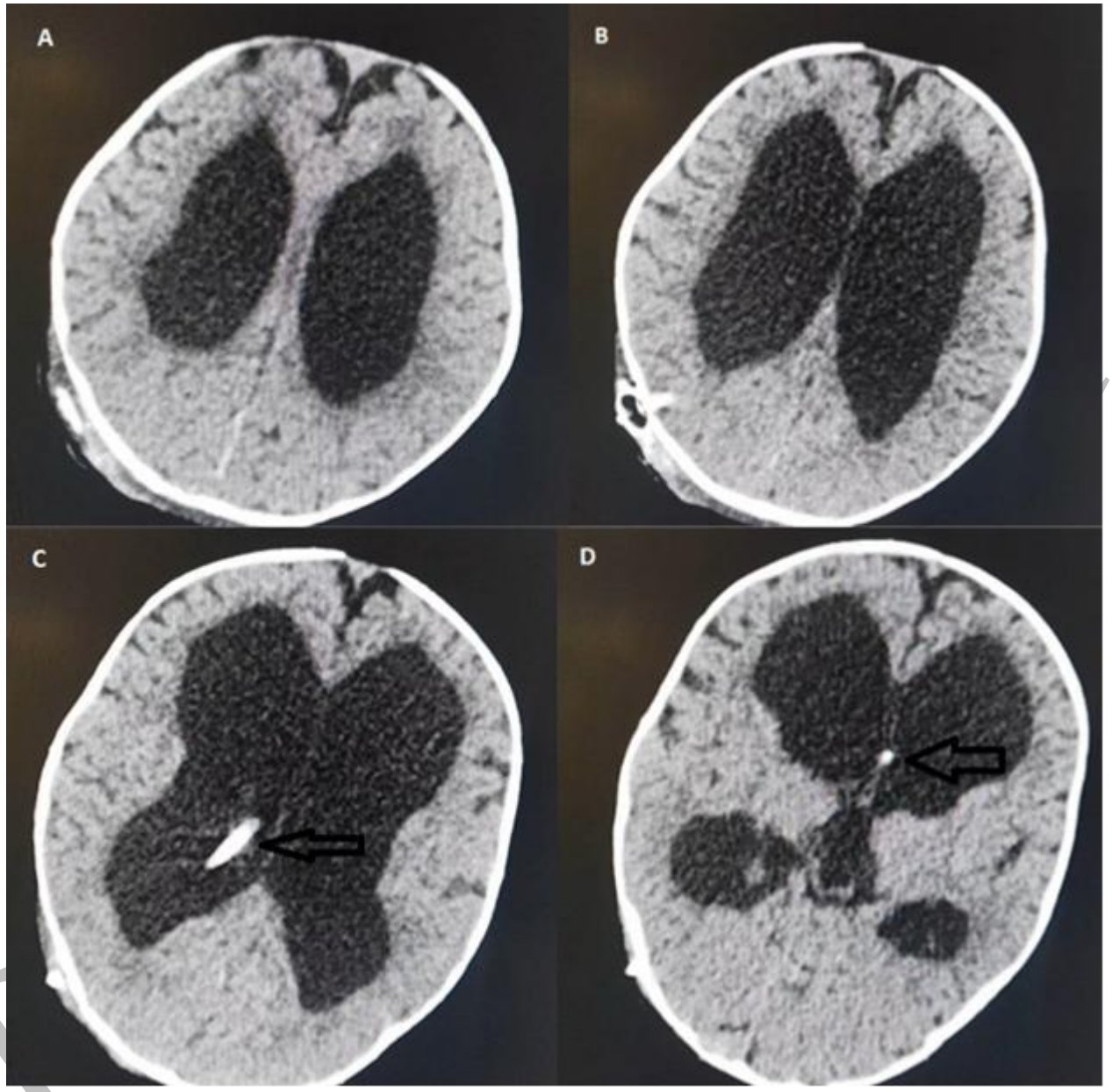

Figure 2. Postoperative native computed tomography of endocranium scan (after ventriculoperitoneal shunt placement): A, B – still dilated lateral ventricles; C – shunt in situ ("arrow"); D – tip of the shunt in left lateral ventricle ("arrow")

