

## Konjenital miyastenya gravisli 2 yaşındaki çocuk hastada kaudal epidural anestezi

Esra Çalışkan, Aysu Koçum, Mesut Şener, Nesrin Bozdoğan, Anış Arıboğan

### ÖZET

Miyastenya gravis nöromusküler kavşakta asetilkolin reseptörlerine karşı antikör oluşumuyla karakterize otoimmün bir hastalıktır. Çeşitli anestezi ajanlarıyla etkileşiminden dolayı anestezi için özel bir ilgi alanı oluşturur. Literatürde miyastenya gravisli çocuk hastalarda erişkin miyastenik hastalardan farklı olarak, anestezi yaklaşımıyla ilgili çok az bilgiye rastlanmıştır ve mevcut bilgilerin çoğunluğu timektomi için yapılan uygulamalarla ilgilidir. Buna ek olarak kaudal anestezi pediatrik hastalarda oldukça yaygın kullanılan bir teknik olmakla beraber miyastenik hastalarda kullanımına ilişkin bir bilgiye rastlanmamıştır.

Biz bu olgu sunumunda 2 yaş, 12 kg ağırlığında miyastenya gravisli erkek hastada kaudal anestezi uygulamasını sunduk. Orşiopeksi ameliyatı planlanan hastada konjenital miyastenya gravis nedeniyle jeneralize güçsüzlük, solunum sıkıntısı ve göz kapaklarında düşüklük mevcuttu. Operasyon odasında rutin monitorizasyonu takiben hasta 1mg midazolam ve 10 mg ketamin ile sedatize edilerek, 1 mL.kg-1 % 0.25 bupivakain ile kaudal blok yapıldı. Lokal anestezi enjeksiyonunu takiben 17 dakika sonra operasyon başladı ve 45 dakika sürdü. Hastada intraoperative ek analjezik ihtiyacı olmadı ve postoperatif takip sorunsuz geçti.

Miyastenya gravisli hastalarda respiratuar ve istemli kas aktivitesinin korunmasına özel bir önem gösterilmelidir. Kaudal anestezi miyastenya gravisli hastalarda kas gevşetici ve endotrakeal entübasyon olmadan havayolunun kontrolüne izin verir.

Sonuç olarak, biz kaudal anestezi miyastenik pediatrik hastalarda uygun ve güvenli bir teknik olduğunu ve bu hastalarda genel anesteziye alternatif olarak uygulanabileceğini düşünüyoruz.

**Anahtar Kelimeler:** Miyastenya gravis, kaudal anestezi, pediatrik cerrahi, anestezi yönetimi

### SUMMARY

#### **Caudal epidural anesthesia for a 2-year old child with congenital myasthenia gravis**

*Myasthenia gravis is an autoimmune disease with antibodies directed against the acetylcholine receptor at the neuromuscular junction. Anesthetists have a special interest in myasthenia gravis because of its interaction with various anesthetic agents. Unlike adult myasthenic patients; very little report has been written about the anesthetic management in children, other than in relation to thymectomy. Although the use of caudal anesthesia in pediatric patients is common, have not seen any report concerning its use in a myasthenic child.*

*In this case report, we represented a 2 year-old boy was performed caudal anesthesia for orchiopexy operation. He had presented difficulty in breathing, generalized weakness and droopy eyes due to congenital myasthenia gravis. In the operating room, following the routine monitoring, the patient was sedated with intravenous 1mg midazolam and 10 mg ketamine. Then caudal block was performed. 17 minutes later from the local anesthetic injection; operation was started and lasted 45 minutes. The patient did not require intraoperative supplemental analgesia and postoperative course was uneventful.*

*Specific attention should be paid to voluntary and respiratory muscle strength in myasthenia gravis patients. Caudal anesthesia allowed airway control of myasthenia gravis patients without endotracheal intubations and muscle relaxant.*

*In conclusion, we think that caudal anesthetic technique may be considered as a safe and suitable for the myasthenic child and it may represent a valid alternative to general anesthesia for these patients.*

**Key words:** Myasthenia gravis, caudal anesthesia, pediatric surgery, anesthetic management.

Başkent Üniversitesi Tıp Fakültesi Anesteziyoloji ve Reanimasyon Ana Bilim Dalı

#### **Başvuru Adresi:**

Yard. Doç. Dr. Esra Çalışkan  
Başkent Üniversitesi Tıp Fakültesi Anesteziyoloji ve Reanimasyon Ana Bilim Dalı  
e-posta: esra\_ertr@yahoo.com  
Tel.: 0.322 458 68 68 / 1200

Baskent University Faculty of Medicine Department of Anesthesiology and Reanimation

#### **Correspondence to:**

Esra Çalışkan Assoc. Prof.,  
Baskent University Faculty of Medicine Department of Anesthesiology and Reanimation  
e-mail: meorhan@gata.edu.tr  
Tel.: +90.322 458 68 68 / 1200

Başvuru tarihi: 09.04.2008, Kabul tarihi: 15.09.2008

## Introduction

The congenital myasthenias comprise a number of genetically determined diseases showing variable muscle weakness from birth. The incidence of myasthenia gravis has been reported 1: 40.000 in the general population (McClelland 1960) and 1% of cases had congenital type (Dalal et al. 1972). A structural malformation of the post-synaptic membrane, with almost complete absence of functional folds, may lack the fine structural specialization with a consequent reduction in acetyl-choline receptor insertion sites. More commonly, the onset of symptoms is after the neonatal period within the first one to two years of life (Baraka 1992). The course of disease is protracted but the symptoms are mild (Dalal et al. 1972).

The anesthetic management of myasthenia gravis in the adult has been well elucidated. However, very little has been written about the anesthetic management in children, other than in relation to thymectomy (Dalal et al. 1972). As it is the case in adulthood, in children as well, if it is possible, regional anesthesia is preferred because of higher incidence of postoperative respiratory failure due to combination of general anesthesia and muscle weakness. Today, pediatric caudal epidural anesthesia is a well-accepted technique which occasionally may serve as the sole anesthetic method in high risk patients as well (Edler 2003). Although use of spinal anesthesia is recently reported from our clinic in a myasthenic child (Koçum et al. 2008), surprisingly we have not seen any report concerning use of caudal block in a myasthenic child. In this case report, we report the anesthetic management for a child with congenital myasthenia gravis using caudal block under sedation.

## Case Report

A 2 year-old boy, weighing 12 kg required orchiopexy. He had presented difficulty in breathing, generalized weakness and droopy eyes due to congenital myasthenia gravis. His EMG on repetitive stimulation and neostigmin test were confirming the diagnosis of myasthenia gravis. He was taking pyridostigmine 15 mg four times daily. We discussed with the parents and decided use of caudal anesthesia.

On the day of surgery the first dose of pyridostigmine was omitted and no pre-medication

was administered. In the operating room, the following parameters were continuously monitored: 5-channel EKG, pulse oxymeter and non-invasive blood pressure measurement. After intravenous access was secured the patient was sedated with intravenous 1mg midazolam and 10mg ketamine and, given 5 % glucose in 0.3 % saline 5 mL. kg<sup>-1</sup> intravenous for intraoperative fluid maintenance during the surgery. Then caudal block was performed and, 1 mL.kg<sup>-1</sup> bupivacaine 0.25 % was injected into the epidural space with a 22 G Epican Paed (Braun, Melsungen, Germany) caudal needle. A lower extremity motor block was obtained. The degree of motor block was 2 (inability to flex knees) according to modified Bromage scale (Bromage, 1965). The sensory level of block was assessed with a skin prick response and, seemed to be T3-4. Skin incision was started 17 min later from the local anesthetic injection; the operation lasted 45 min and the patient did not require intraoperative supplemental analgesia. He remained comfortable and hemodynamically stable. Intraoperatively, heart rate remained at 84-98 bpm, blood pressure (systolic/diastolic) between 97/108 to 47/62 mm Hg and, oxygen saturation between 99 % and 100 %. The patient recovered smoothly from anesthesia. Motor block had resolved at 2 h completely. The postoperative course was uneventful. Urinary retention was not seen and spontane micturition was returned 2.5 h at postoperative period.

The pain relief medication was provided oral paracetamol every 6 h as required at postoperative period. First analgesics requirement was observed fourth hour after local anesthetic injection. The patient was discharged from the hospital first day after the surgery.

## Discussion

Caudal block with inhalation anesthesia is commonly performed in small children for the procedures below umbilicus for outpatient surgery, combining prolonged analgesia with few side effects. However, myasthenic patients may experience some depression of neuromuscular function with inhalation anesthetics (Abel and Eisenkraft 2002) Specific attention should be paid to voluntary and respiratory muscle strength in these patients. The patient ability to protect and maintain a patent airway postoperatively may be compromised if any bulbar involvement exists preoperatively. The ability to cough and clear secretions may be compromised as well. Alt-

though ketamine increases salivation, airway is usually well maintained with preservation of pharyngeal and laryngeal reflexes during ketamine anaesthesia. In addition to, ketamine is an intravenous anesthetic with analgesic properties in subanesthetic doses (Jahangir et al. 1993). It has been shown that use of a single small dose of ketamine in a pediatric population undergoing tonsillectomy could reduce the frequency or even avoid the use of rescue analgesic in postoperative period (Conceição et al. 2006).

Our patient hasn't got any swallowing problems or symptoms of bulbar involvement. So, we prefer ketamine and low dose of midazolam rather than inhalation anesthesia to maintain respiration and provide additive postoperative analgesic effect in order to better tolerance of caudal anesthesia.

The most important two variables that determine the safety of caudal block are type of local anesthetics and its volume in myasthenic patients. Local anesthetics agents decrease the sensitivity of the post-junctional membrane to acetylcholine and the use except for therapeutic drug dosages of these agents might cause weakness in myasthenic patients. Ester anesthetics, which are metabolized by cholinesterase, may present particular problems in patients taking anticholinesterase drugs. Regional and local anesthesia should be performed using reduced doses of amide (rather than ester) local anesthetics to avoid high blood levels and also to avoid blockade of the innervations of intercostal muscles to minimize the risk of respiratory muscle weakness (Abel and Eisenkraft 2002). Because of its long duration of action, bupivacaine is the local anesthetic agent used most commonly for caudal epidural blockade. We used bupivacaine for our myasthenic patient as it's an amide type local anesthetic and it has long term opioid sparing effect by continuing analgesia into the postoperative period.

The management of postoperative pain may be an important problem in myasthenic patients. The choices of analgesic agent and route of administration are important because postoperative pain may compromise pulmonary function which is already impaired by myasthenia. Recently, the safe and successful thoracic epidural anesthesia with bupivacaine for intraoperative anesthesia and postoperative analgesia for

transsternal tymectomy in adult myasthenic patients has been reported (Akpolat et al. 1997). Opioid analgesics in therapeutic concentration do not appear to depress neuromuscular transmission in myasthenic muscle but, central respiratory depression may be a problem with opioids. However, intraoperative and postoperative analgesia was described with parenteral opioid and continuous patient controlled analgesia techniques respectively for tymectomy (White and Stoddart 2004). Remifentanyl has been used by Lorimer et al and is beneficial as it blunts airway reflexes and reduces respiratory drive (Lorimer and Hall 1998). We prefer ketamine as additive analgesic agent to caudal anesthesia in our patient.

Several formulas have been described to determine dosages and volumes required to produce blockade to various levels. Dalens reported excessive spread of local anaesthetic in 30% of patients receiving 1.25 ml/kg and found 0.75-1 ml/kg to be appropriate for a wide variety of operations (Dalens and Hasnaoui 1989). The use of 0.75 ml/kg 0.25% bupivacaine for both hernia repair and orchidopexy resulted in 69% of children being pain free 4 h postoperatively (Fisher et al. 1993). Pharmacokinetic data from several studies show that single epidural doses of 2-2.5 mg/kg of bupivacaine are associated with low plasma levels of bupivacaine. A simple working rule is for children less than 6 months of age is that 1 ml/kg of bupivacaine 0.125 % will block low thoracic dermatomes. In children above this age 1 ml/kg of bupivacaine 0.25 % will consistently block inguinal dermatomes in children less than 20 kg while above this weight the technique becomes inconsistent at blocking inguinal dermatomes (Rowney and Doyle 1998). Based on previous data mentioned above, we aimed to use such a minimal dose of bupivacaine which may effectively block inguinal dermatomes. We obtained effective surgical anesthesia and postoperative analgesia without any requirement for opioid analgesics for postoperative 3 hours with administering 1 ml/kg 0.25 % bupivacaine (nearly 2.5 mg/kg of bupivacaine) caudally combined with intravenous ketamine and midazolam.

Spinal anesthesia has the advantage of reduced drug dosage; it is also an effective alternative to caudal block whereas caudal block procedure may provide a longer duration of anesthesia than spinal anesthesia as well as postoperative anal-

gesia (Rowney and Doyle 1998).

In summary, we conclude that caudal anesthesia may be effective and suitable anesthetic technique for the myasthenic child provided that a proper local anesthetic agent is chosen with a proper dose.

## References

- Abel M, Eisenkraft J.B. Anesthetic Implications of Myasthenia Gravis. *Mt Sinai J Med.* 2002 ;69(1-2):31-7.
- Akpolat N, Tilgen H, Gürsoy F, Saydam S, Gürel A. Thoracic epidural anaesthesia and analgesia with bupivacaine for transsternal thymectomy for myasthenia gravis. *Eur J Anaesthesiol* 1997; 14:220-223.
- Baraka A. Anesthesia and myasthenia gravis. *Middle East J Anesthesiol.* 1992; 39(5):476-86.
- Bromage FR. A comparison of the hydrochloride and carbon dioxide salt of lidocaine and Prilocaine in epidural analgesia. *Acta Anaesthesiol Scand* 1965 (Sp.XVI): 55-69.
- Conceição MJ, Bruggemann DA, Conceição D, Carneiro LC. Effect of intravenous single dose of ketamine on postoperative pain in tonsillectomy patients. *Paediatr Anaesth* 2006; 16(9):962-7.
- Dalal F.Y, Bennett E.J, Gegg W.S. Congenital myasthenia gravis and minor surgical procedures. *Anaesthesia* 1972;27(1):61-65.
- Dalens B, Hasnaoui A. Caudal anesthesia in pediatric surgery: success rate and adverse effects in 750 consecutive patients. *Anesth Analg* 1989; 68:83-9.
- Edler A. Caudal epidural anesthesia for pediatric patients: A safe, reliable and effective method in developing countries. *World Anesthesia* 2003;17 (article 5)
- Fisher QA, McComiskey CM, Hill JL, Spurrier EA, Voigt RE, Savarese AM, Beaver BL, Boltz MG. Postoperative voiding interval and duration of analgesia following peripheral or caudal nerve blocks in children.
- Jahangir SM, Islam F, Aziz L. Ketamine infusion for postoperative analgesia in asthmatics: a Comparison with intermittent meperidine. *Anesth Analg* 1993; 76:45-9.
- Kocum A, Sener M, Bozdogan N, Turkoz A, Arslan G. Spinal anesthesia for inguinal hernia repair in 8- year old child with myasthenia gravis. *Paediatr Anaesth* 2007; 17(12): 1220-1.
- Lorimer M, Hall R. Remifentanyl and propofol total intravenous anaesthesia for thymectomy in myasthenia gravis. *Anaesth Intensive Care* 1998; 26(2):210-2.
- McClelland R.M.A. The myasthenic state and the myotonic syndrome. *British J of Anaesth* 1960; 32:81
- Rowney D.A, Doyle E. Epidural and subaracnoid blockade in children. *Anaesthesia* 1998,53:980-1001.
- White MC, Stoddart PA. Anesthesia for thymectomy in children with myasthenia gravis. *Paediatr Anaesth.* 2004; 14(8):625-35.