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ANESTHESIA FOR CESAREAN SECTION IN A PATIENT WITH SPINAL MUSCULAR ATROPHY *Habib, A.S. Helsley, S.; Millar, S.; Muir, H.A. Anesthesiology, Duke University Medical Center, Durham, NC*

Introduction: Proximal spinal muscular atrophies (SMA) are a group of inherited neuromuscular disorders characterized by degeneration of the anterior horn cells. We report the anesthetic management for cesarean section (CS) of a patient with type II SMA. Case report: A 23 years old primiparous woman, with Werndig-Hoffman type II SMA, was scheduled for an elective CS at 36 weeks gestation secondary to inadequate pelvis. Pregnancy was complicated by an attack of pneumonia at 28 weeks treated with antibiotics and oxygen therapy. Her SMA was diagnosed at13 months of age. She never walked and has been wheelchair bound. Severe kyphoscoliosis necessitated 5 back surgeries. Overnight ventilatory support was needed following the last surgery. The investigations were significant for severe restrictive lung disease (FVC =0.86 l, FEV1 = 0.78 l), PaO2= 77mmHg, PaCO2=38 mmHg, bicarbonate=28mmol/l and SaO2=93 %. A back x-ray revealed rods extending from the upper thoracic to the sacral area. She was 1.25m.tall and weighed 37 kg. Her airway was Mallampati class III, hyomental distance 3 fingers, good mouth opening and neck movement. Musculoskeletal examination revealed severe kyphoscoliosis, pectus excavatum, slender limbs with severe contractures and 2-3/5 motor strength. Following administration of sodium citrate, application of routine monitors and preoxygenation, anesthesia was induced using alfentanil 800 mcg, propofol 150mg and lidocaine 40 mg with cricoid pressure. A size 6 ETT was easily inserted. A radial arterial line was placed. Maintenance was with N2O in O2 and isoflurane. 10U oxytocin were given following delivery. Analgesia was provided using alfentanil 1500 mcg and morphine 6 mg with ketorolac 30 mg and an ilioinguinal block at the end of surgery. A viable male infant weighing 2030 gm (Apgar scores 2,9) was delivered The patient was extubated and sent to the intensive care unit (ICU). She was discharged home after 5 days. Discussion: We chose general anesthesia since the patient had 5 previous back surgeries with extensive scarring and Harrington rods extending to the sacral area. Her previous general anesthetics were uneventful with no documented difficult intubation or malignant hyperthermia reaction. We avoided the use of suxamethonium due to the risk of causing hyperkalemia and rhabdomyolysis. These patients may also have increased sensitivity to non-depolarizing muscle relaxants. The use of the Propofol and alfentanil technique avoided the use of muscle relaxants and provided good intubating conditions. An arterial line was inserted for frequent blood gas sampling. The patient was nursed in ICU since autotransfusion may precipitate cardiorespiratory decompensation. Ketorolac was given and an Ilioinguinal block was performed to reduce opiate requirements.

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ANESTHETIC MANAGEMENT FOR DELIVERY FOR A PARTURIENT WITH MAY-HEGGLIN ANOMALY: A CASE REPORT *Calinaran, A.L. Wong, C.A. Northwestern University Medical School, Chicago, IL*

We describe the anesthetic management of a parturient with May-Hegglin anomaly, a rare hereditary platelet abnormality. A G_4P_0, 28-year-old with May-Hegglin anomaly was referred for pre-anesthetic evaluation at 36 w gestation. Diagnosis was made when incidental thrombocytopenia was found at 11 y of age. Her medical history included 3 spontaneous abortions, and a D&C with no abnormal bleeding despite a platelet count (PC) of $<20{,}000/mm^3$. She was subsequently diagnosed as a heterozygote carrier of the Leiden Factor V mutation. A hematology consultation noted the lack of clinical signs of bleeding, with PC of $23{,}000/mm^3$; bleeding time 2 min (normal < 9 min); platelet function analysis (PFA) to epinephrine 238 s (normal 58-171 s) and ADP 107 s (normal 41-134 s). Her anesthesiologists counseled her of the uncertain risk of spinal hematoma after neuraxial blockade. Because she was unwilling to undergo labor without neuraxial analgesia, and the potential risk of fetal intracranial hemorrhage with vaginal delivery, a decision was made to proceed with elective cesarean delivery. On the day of surgery her physical examination was WNL; her PC was $18{,}000/mm^3$ and Hct 40%. Preoperative platelet transfusion was not given because of her previous uneventful D&C and negative bleeding history. She had an uneventful general anesthetic and operative procedure with estimated blood loss of 800 mL. Her postoperative course was unremarkable. Tests done on the infant showed May-Hegglin anomaly. The majority of patients with May-Hegglin anomaly are asymptomatic but some present with bleeding abnormalities. Laboratory findings include thrombocytopenia, giant platelets, and inclusion bodies in leukocytes. The abnormal PFA results may have resulted from severe thrombocytopenia as this is known to affect PFA values. Also of interest in this case was the Leiden Factor V mutation, a condition associated with hypercoagulation. Indeed, her history of 3 spontaneous abortions supports a hypercoagulable state, as this mutation is associated with an increased risk of abortions (1). We elected to perform general, rather than spinal anesthesia for cesarean delivery because of the unknown risk of spinal hematoma. Two previous case reports on the management of parturients with May-Hegglin anomaly describe the uneventful use of spinal anesthesia (2). However, the baseline platelet counts were higher and the parturients received prophylactic platelet transfusions. We therefore recommend that anesthetic options be thoroughly discussed with the parturient prior to expected delivery, and close involvement of the anesthesiologist, obstetrician, and hematologist in the care of patients affected with this disorder. *1) N Eng J Med 2000;343:1015-8 2) J Reprod Med 1993;38:311*