Case Report: Seckel’s Syndrome and anesthesia: a case report

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Summary: Seckel’s syndrome (SS) is a rare form of proportional microcephalic primordial dwarfism characterized by a “bird-headed” appearance. The literature includes only two case reports regarding the anesthetic management of this condition (1,2). We report two anesthetics on a single pediatric patient with Seckel’s syndrome complicated by bilateral moyamoya syndrome.

Case Report: A 16 year-old girl with SS, weighing 13 kg, presented with a history of headaches and TIAs of her left face and hand over the past year. MRI showed multiple deep white matter infarctions. An MRA diagnosed bilateral moyamoya syndrome. She was evaluated with angiography under general anesthesia (GA) and underwent a second GA for bilateral pial synangiosis surgery. Each anesthetic included IV prehydration, 1 mg Midazolam IV, induction with either Diprivan or Thiopental and nondepolarizing muscle relaxant facilitating easy mask ventilation. Her vocal cords were easily visualized and intubated with 4.5 low profile cuffed endotrachial tube. Anesthetic maintenance included Forane vapor and Fentanyl. Normocapnea and normotension were maintained throughout. She was extubated in the OR on each occasion without event and discharged home without complication five days following her craniotomy.

Discussion: Seckel’s syndrome is an autosomal recessive dwarfism with an incidence of less than 1 in 10,000 cases (1). Patients are characterized by intrauterine growth retardation, microcephaly, beak-like nose, micrognathia and variable mental retardation. Fewer than 60 cases ave been reported. Our case is the 7th report of a CVA in an SS patient (3 moyamoya, 4 aneurysms) (3). Moyamoya is an idiopathic intracranial internal carotid artery stenosis that results in ischemic infarcts in children. The surgical and anesthetic management for moyamoya syndrome have been reviewed elsewhere (4). Although retrognathia is common in SS, there are no documented cases of airway obstruction or difficult intubation. Postop apnea in a 5 year-old and subglottic stenosis in an adult have been reported (1,2). Dentition matures normally, but teeth are malpositioned and crowded (5). The cervical spine is unaffected. Our patient had undergone several GAs prior to her presentation without reported airway difficulty. Endotrachial tube size would seem to be better predicted by the patient’s weight, which holds true for our case (1). These patients typically have small, friable veins that are difficult to cannulate (1). An instability on chromosome 3, at the same locus that causes ataxia-telangiectasia, is responsible for SS, suggesting a cause for the reports of myelodysplasia and acute myeloid leukemia seen in SS (6). A recent blood count is recommended prior to surgery. Cases of malignant hypertension leading to a cerebral hemorrhage and benign periop hypertension have been reported by authors who suggest short stature as the cause of hypertension (2,7).

Conclusion: The anesthetic of a patient with SS is reminiscent of that of a neonate. Care should be taken during the preoperative evaluation highlighting the airway, neurological status and hematological system.

References: