Case Description: A 19-year-old, male patient weighing 58 kg was scheduled for an elective D1–D2 laminectomy and excision of an arachnoid cyst under general anesthesia. Diagnosed with tricuspid atresia at birth, he underwent modified right Blalock–Taussig shunt as a neonate and a hemi-Fontan procedure at 8 months of age. He was NYHA class 2 until 1 month ago until he developed muscle weakness with grade-4 power. The ECHO done preoperatively showed functional hemi-Fontan's shunt, restrictive VSD 2 mm, small ASD, rudimentary RV, dilated LV, and a normal LV systolic function. After establishing standard ASA monitors, induction, and tracheal intubation were performed with intravenous fentanyl, ketamine, etomidate, and atracurium. Anesthesia was maintained with sevoflurane, MAC 0.5 and propofol infusion titrated with BIS. The patient was positioned carefully in prone with transcutaneous pacing pads and MEP was monitored intraoperatively to guide excision.

Conclusion: Understanding the Fontan physiology, the single ventricle physiology and pulmonary blood flow is critical to maintain hemodynamic stability. Anesthetic management, enabling intraoperative MEP monitoring, while maintaining cardiac function in the setting of altered cardiac physiology is essential for successful management.

A033 Craniotomy in a Case of Eisenmenger's Syndrome with Pulmonary Artery Hypertension: A Neuroanesthetic Challenge

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Background: The Eisenmenger syndrome is a form of cyanotic congenital heart disease not amenable to corrective surgery. It develops in patients with left-to-right shunts that results in concomitant pulmonary hypertension and right heart volume overload. The reported perioperative mortality of these critically ill patients is as high as 20 to 30%.

Case Description: An 18-year-old female patient presented to our institute for neurosurgical management of third ventricular colloid cyst. She was a diagnosed case of double-outlet right ventricle (DORV) with patent ductus arteriosus (PDA) with hypoplastic left aortic arch with pulmonary arterial hypertension (PAH) and Eisenmenger syndrome. She underwent right frontal craniotomy and tumor decompression under general anesthesia. She was managed with the hemodynamic goals to avoid any decrease in preload, decrease in systemic vascular resistance, increase in pulmonary vascular resistance, and decrease in myocardial contractility.

Conclusion: We would like to discuss the unique set of challenges encountered in perioperative management of these critically ill patients with reference to the present guidelines available and share how we managed this case.

A034 Anesthetic Management of a Patient of Pituitary Microadenoma with Low Ejection Fraction for Inferior Petrosal Sinus Sampling

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Background: Adrenocorticotropic hormone secreting pituitary adenoma is though quite rare yet associated with increased morbidity and mortality due to severe comorbidities associated with them. Inferior petrosal sinus sampling (IPSS) is the gold standard invasive procedure for the anatomical localization for Cushing's disease.

Case Description: We report two typical cases of Cushing's disease with low–ejection fraction who underwent IPSS and discuss the difficulties encountered and their management. Both of them were female patients of age 40 and 38 years, respectively. They presented with similar complaints of weight gain, facial hair, and body ache and after clinical examination and investigations, they were diagnosed to have Cushing's disease with pituitary microadenoma. Hypertension and diabetes mellitus were present with echo findings of ejection fraction of 30 and 35%, respectively. IPSS was performed under general anesthesia. Patients were hemodynamically stable throughout the procedure and we avoided any tachycardia, hypoxia, hypercarbia, hypothermia, and proper analgesia was ensured. Blood sugar levels were kept within normal limits.

Conclusion: Anesthetic implications due to Cushing's disease include hyperglycemia, hypertension, proximal muscle weakness, and skin thinning. Delicate skin and osteoporosis cause difficulty in venous access and increases risk of spontaneous fractures. A thorough understanding of preoperative assessment of airway, neurological, and endocrine status were helpful for our management.

A035 Craniotomy in Klippel–Trenaunay Syndrome: Concerns and Challenges

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Background: Klippel–Trenaunay Syndrome (KTS) is a rare congenital disorder characterized by cutaneous capillary malformations, soft tissue and bone hypertrophy, and venous varicosities. Multisystem involvement of this syndrome mandates adequate preparation and planning, with meticulous conduct of anesthesia, and surgery in these patients to achieve favorable outcomes.

Case Description: A 17-year-old girl presented for excision of right frontal calvarial hemangioma measuring 8 cm × 9 cm × 5 cm with intracranial extension and associated

Chiari-1 malformation. On examination, her neurocutaneous markers suggested classical KTS-presence of distinct, linear bordered port-wine stains. She had lymphoedema of bilateral lower limbs and disproportionately enlarged feet with club feet deformity. Solid organ involvement and congenital heart disease were ruled out. Adequate blood products were arranged in view of anticipated blood loss. Standard anesthetic induction was performed. In view of Chiari malformation and anticipated soft tissue changes in the airway, intubation was performed using C-MAC videolaryngoscope. Patient was positioned carefully and surgery was performed uneventfully.

Conclusion: Patients with KTS pose challenges during vascular access necessitating use of ultrasonography to diagnose and avoid inadvertent injury to superficial arteriovenous fistulous malformations. Similar concerns exist during scalp block regarding inadvertent local anesthetic injection into occult subcutaneous scalp hemangiomas. Presence of associated Chiari-1 malformation and proneness for pathological fractures requires care during intubation and positioning. Oropharyngeal neurofibromatosis lesions warrant preoperative airway examination using indirect laryngoscopy to understand the potential airway difficulty and formulate intubation plan. Calvarial hyperostosis can result in difficulty during craniotomy. Proximity of the hemangioma to the superior sagittal sinus places the patient at risk of venous air embolism.

A036 Role of NIRS in a Child with Moyamoya Disease for Encephaloduroarteriomyosynangiosis (EDAMS): A Case Report

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Background: Moyamoya, "puff of smoke" disease (MMD), is a rare cerebrovascular disease with progressive stenosis of intracranial blood vessels. Anesthetic goal of revascularization surgery is to maintain the cerebral oxygen supply and demand. We wanted to report a case of NIRS-based anesthetic management of encephaloduroarteriomyosynangiosis (EDAMS).

Case Description: An 8-year-old boy weighing 24 kg presented with two episodes of transient ischemic attacks (TIAs), diagnosed with MMD, planned for EDAMS. Overnight intravenous fluid (IVF) was given to prevent dehydration and diazepam premedication was given for anxiolysis. In the operating room, with ECG, EtCO2, SPO2, NIRS, and NIBP monitoring were attached; induction and intubation were performed using propofol, fentanyl, and atracurium. Postintubation, arterial BP, temperature, and urine output monitoring was established. Regional scalp block with 0.2% ropivacaine was given for analgesia. Anesthesia was maintained using air, oxygen, and sevoflurane. Depth of anesthesia, brain oxygenation, and hemoglobin trends were monitored using the patient state index, NIRS, and noninvasive hemoglobin analyzer using SedLine, Masimo. MAP, CO2, temperature, and depth of anesthesia were tightly controlled to keep the cerebral oxygenation close to the baseline. Repeat regional scalp block, fentanyl infusion was given for postoperative analgesia. He was discharged on the POD 4 without neurological deficit.

Conclusion: MMD has limited cerebrovascular reserve with propensity to develop cerebral ischemia during the perioperative period. In our case, the balance between the cerebral oxygen supply and demand was strictly maintained by monitoring the cerebral oxygenation using NIRS. Various factors, such as MAP, EtCO2, temperature, depth of anesthesia, and intraoperative hematocrit were controlled within normal limit to maintain the cerebral oxygenation close to baseline throughout surgery which aided for the successful outcome.

A037 Utility of Compass 31 Questionnaire to Predict the Autonomic Dysfunction and Intraoperative Hemodynamic Fluctuations and Correlate it with CASS Score in Patients with Compressive Cervical Myelopathy

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Background: To assess utility of COMPASS-31 questionnaire as screening tool for diagnosing autonomic dysfunction (AND) and correlate it with modified CASS score to predict the severity of AND, intraoperative hemodynamic fluctuations in patients undergoing surgery for compressive cervical myelopathy (CCM).

Materials and Methods: After obtaining IRB approval and informed consent, 42 ASA I-II patients, aged 18 to 70 years, who underwent decompression for CCM were recruited. In the preoperative period, COMPASS 31 questionnaire was given in the language they could comprehend, and the total score was calculated. They underwent autonomic function tests and modified CASS score was calculated. During intraoperative period, standard anesthesia protocol followed and hemodynamics were noted at regular intervals throughout the surgery. Ephedrine/phenylephrine boluses or noradrenaline infusion was given to maintain a target BP within 20% of baseline. Amount of vasopressor used was recorded.

Results: The mean age of the patients studied was 48.76 ± 10.69 years. Fifty percent of patients had Nurick's grades 2 and 3, and the rest had Nurick's grades 4 and 5. All patients had AND varying from mild (46%), moderate (12%) to severe (42.9%). The median COMPASS-31 score (IQR: 25–75%) was 19 (6–33). Receiver operating characteristic (ROC) analysis revealed COMPASS-31 had fair accuracy with area under the curve (AUC) = 0.738 (p = 0.009). Total COMPASS-31 score of 30 had sensitivity of 52.2% and specificity of 89.5% to detect moderate to severe AND. The median (IQR: 25–75%) Modified CASS score was 3 (2–5). Moderately positive correlation 0.383 (p = 0.05) found between CASS and COMPASS-31 score. Nurick's grade showed positive correlation with CASS/COMPASS-31with correlation coefficient of 0.35 (p = 0.023), 0.48 (p = 0.001), respectively. In patients with severe AND had significant hemodynamic fluctuation compared with mild/moderate AND.