Anaesthesia for Neurosurgical Procedures in Neonates

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40.1 Introduction

A newborn can present with several conditions affecting the central nervous system (CNS) and involving the brain and spinal cord. Common neurological conditions are seizures, birth asphyxia, encephalopathy, hypotonia, intracranial hemorrhage (ICH), hydrocephalus, cerebrovascular (CV) malformations, congenital brain tumours, neural tube defects (NTDs), and Arnold Chiari malformations (ACMs). Some of these such as hydrocephalus, ICH, NTDs, ACMs, CV malformations, and tumours are amenable to surgical treatment.

Undertaking neurosurgery in neonates presents a set of major challenges to the neurosurgeon, anaesthesiologist, and the neonatologist providing pre- and post-surgical care. The surgeries are mostly high-end and the neonatal population precarious and delicate. Neonates have unique multi-system anatomical and physiological attributes and concomitant congenital anomalies. The neonatal CNS is particularly vulnerable to perioperative injury due to a still-developing brain and spinal cord, and a very different and evolving neurophysiology that is responsible for the variable and unpredictable responses to surgery and anaesthesia. The variety of neurosurgical lesions, each with a distinct pathophysiology, clinical manifestations, and management techniques, add to the challenges. Considerations of the interrelated pathophysiologic processes in neonates are thus important to fully understand the nature of the neurological abnormality and its effective treatment which would enable a successful interdisciplinary management of neurosurgery in neonates.

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40.2 Neonatal Neurology

40.2.1 Cerebral Blood Flow and Perfusion Pressure

A very different cerebrovascular physiology and cranial bone development distinguishes neonates from older children and adults. An adequate cerebral perfusion pressure (CPP) is vital for delivery of oxygen, glucose, and other vital nutrients to the brain. Any perturbation in the cerebral blood flow (CBF) can lead to hypoxic brain injury and ICH. CBF in neonates is tightly coupled with cerebral metabolic oxygen ($CMRO_2$) requirements, and both increase proportionally immediately after birth. Based on the principle of flow-metabolism coupling, neonates have a lower $CMRO_2$ (2.3 mL/100 g/min) compared to adults (3.5 mL/100 g/min) and consequently a lower CBF (40–42 mL/100 g/min) than in adults (55 mL/100 g/min). Due to the fewer synaptic connections and a lower neuronal activity in the neonatal brain, neonates have a lower oxygen (O_2) requirement and a relative tolerance to hypoxemia. $CMRO_2$ increases to above adult values in infancy and older children to 5.2 mL/100 g/min with consequent increase in CBF to 90 mL/100 g/min at age of 6 months−3 years, and 100 mL/100 g/min between 3 and 12 years of age [1, 2].

The cerebral vascular response to $PaCO_2$ is poorly developed in neonates. Hypercapnia is a potent cerebral vasodilator in adults with linear increase in CBF between $PaCO_2$ of 20 and 80 mmHg, and hypocapnia, potent cerebral vasoconstrictor. The neonatal cerebral vasculature response to $PaCO_2$ is poorly developed. There is significant increase in total CBF in response to hypercarbia, but neonates are relatively immune to hypocapnia. They respond with lesser cerebral vasoconstriction to moderate hypocapnia ($PaCO_2 > 15$ mmHg) with maintenance of CBF. But, severe hypocapnia ($PaCO_2 < 15$ mmHg) results in decrease in CBF and increase in heart rate.

Contrarily, the response to PaO_2 is more pronounced in neonates compared to adults. The CBF in neonates increases even with a small decrease in PaO_2, while in adults it increases only when the PaO_2 falls below 50–60 mmHg. This heightened response in neonates can be attributed to their relative hypoxic state (normal PaO_2 is 65–70 mmHg in neonates vs 95–100 mmHg in adults) and increased affinity of O_2 to fetal haemoglobin (fHb) [2, 3].

40.2.2 Cerebral Autoregulation

Cerebral autoregulation is a vital protective mechanism controlling cerebral circulation with changes in mean arterial pressure (MAP) and extends from MAP of 50 to 150 mmHg in adults. The cerebral autoregulation limits in neonate vary with lower cerebrovascular reserve and are difficult to ascertain, but the lower limit of MAP of 40 mmHg is considered the critical CPP in neonates. While cerebral autoregulation may be intact in healthy term neonates, premature with low birth weight have a linear correlation between CBF. Extreme rise of blood pressure in absence of

cerebrovascular autoregulation and fragile vasculature predisposes neonates to intraventricular hemorrhage (IVH), more so in a preterm neonate. Hence, there is a need of a tight blood pressure control during anaesthetic management to minimize both cerebral ischemia and risk of IVH [2, 4–6].

The percentage of cardiac output (CO) directed to the brain differs in neonates; CBF is 10–20% of CO during the first 6 months. The head of the neonate accounts for a large percentage of the body surface area and blood volume which places the neonate at risk for significant haemodynamic instability during neurosurgical procedures [7, 8].

40.2.3 Intracranial Pressure (ICP)

ICP also depends on age. It is 0–6 mmHg in infants, 6–11 mmHg in toddlers, and 13–15 mmHg in adolescents and adults. The newborn experiences physiological salt and water loss and consequent weight loss in the first 10 days after birth. There is a parallel decrease in cerebral volume and hence fall in ICP that can reach subatmospheric values along with overriding of membranous bones of the skull. Therefore, normal ICP values in neonates are not only lower than in adults, but also differ in the same neonate through the first month. The decrease in ICP during first 10 days can also lead to compensatory ventricular enlargement in accordance with Monroe Kelly Doctrine and lead to what is known as a physiological hydrocephalus that is usually mild. Drop in ICP and development of physiological hydrocephalus can mask diagnosis of a developing pathological hydrocephalus. Hence, hydrocephalus in neonates can be present even with ICP that is seemingly normal if adult standards are used; this is known as **normal pressure infantile hydrocephalus**. However, ICP is high if age-appropriate standards are used. Knowledge of age-appropriate ICP physiology and range allows for early detection and management of hydrocephalus in neonates [9].

The skull is a closed cavity with three compartments; the brain, blood, and cerebrospinal fluid (CSF) (Monroe-Kellie doctrine). An increase in volume of either of these three compartments causes increase in ICP unless compensated by reduction in other two, in adult brain. However, the neonatal cranial vault is in a state of flux. Open fontanels and cranial sutures result in a compliant intracranial space. Skull has open sutures and fontanelle, which not only provide locus for ICP monitoring and trans fontanelle ultrasound (USG), but also for slow increases in intracerebral volume. The mass effect of a slow-growing tumour or insidious haemorrhage is often masked by a compensatory distended fontanel and widening of the cranial sutures, increase in skull volume with minimal increase in ICP. However, acute increases in cranial volume due to massive IVH or an obstructed ventricular system cannot be attenuated by expansion of the immature cranial vault and often result in life-threatening intracranial hypertension. The closure of posterior fontanelle occurs by 6 months, of anterior fontanelle by 12–18 months, and of final cranial sutures by 10 years of age [2, 10].

40.2.4 Blood-Brain Barrier

Blood-brain barrier in neonate is immature and weak, permitting easy permeability of anaesthetic drugs, and thus increased sensitivity. Presence of increased amount of CSF coupled with immature myelination can shorten and decrease the potency of local anaesthetics in the CSF.

40.2.5 Spinal Cord

Spinal cord in neonates extends till L3, two segments below the adult level (L1) and the dural sac ends at S4 (S2 in adults).

Development of CNS is incomplete at birth and continues up to the age of 1 year and poses challenges to the anaesthesiologist for maintaining cerebral oxygenation and perfusion and preventing cerebral injury during neurosurgery [11]. The unique pathophysiologic mechanisms in neonates greatly impact the conduct and outcome of neurosurgical interventions. The biomechanical nature of their brain tissue, distensible skull, and widening sutures results in an abnormal head growth and resultant craniocephalic disproportions that lead to considerable neurosurgical difficulty. Congenital anomalies like open NTDs, despite timely intervention, may still result in neurological deficits due to presence of concurrent microscopic abnormalities like widespread synaptic miswirings which are not correctable by surgery. The unique plasticity of the neonatal CNS tissue determines its vulnerability to neurosurgical injury and its ability to its functional recovery, and hence, estimating prognosis after neurosurgery becomes extremely difficult in this age [12, 13].

40.3 Neurosurgical Conditions in the Neonate

A newborn can present with several neurological conditions attributable to congenital defects or premature or difficult delivery. Disorders like neonatal seizures, newborn stroke, encephalopathy, birth asphyxia, CNS infections, and congenital neuromuscular diseases including myasthenia and muscular dystrophies are mostly managed by medical treatment. Pathological conditions in neonates amenable to surgical treatment include hydrocephalus; NTD [meningomyelocele (MMC), encephalocele, spina bifida, syringomyelia, and tethered cord]; congenital brain and spine malformations like ACMs and arachnoid cysts; brain and spinal cord tumours; CV conditions (Vein of Galen malformation); traumatic brain and spinal cord injuries; craniosynostosis, craniofacial syndromes; and craniopagus conjunctions.

However, neurosurgery in neonates is generally avoided considering their frailty and increased perioperative morbidity and mortality due to a higher risk of developing cardiac and respiratory complications [14]. Hence, non-emergent

surgical conditions are often deferred until after 3 months of age when surgical intervention is medically safer.

Neurosurgeries that may need to be undertaken in neonates, often as urgent interventions, include:

- Drainage or diversion of cerebrospinal fluid (CSF) and intracranial cysts.
- Closure of NTDs like MMCs and encephaloceles.
- Removal of neoplasms, anomalous masses, and haematomas secondary to brain and spinal cord trauma.
- Decompression of type-2 ACMs.
- Opening of bony fusions in craniosynostoses.

Sometimes, two or more of these surgeries may be necessary [12, 15].

Congenital CNS tumours include teratomas, astrocytomas, choroid plexus papillomas, glioblastomas, gangliogliomas, primary neuroblastomas, craniopharyngiomas, and embryonal, ependymal, and meningeal tumours. In view of their large size, mass effect, vascularity, and in context of a patient with low total blood volume, these tumours present formidable surgical and anaesthetic challenges. Malignant brain tumours are the second commonest cancers in children and most occur in the posterior fossa. Surgical resection is the primary treatment modality [16, 17]. Other rare neurosurgical conditions in a neonate are craniosynostosis, arterivenous (AV) malformations, and craniopagus twins. However, these conditions are seldom operated in the neonatal period. Preoperative embolization of choroid plexus tumours helps reduce intraoperative blood loss and protects the vulnerable neonate from haemodynamic instability [18, 19].

Neonatal head trauma may occur due to the birth process resulting in epidural, subdural, subarachnoid, and parenchymal hemorrhages that may require percutaneous aspiration of blood. A high head to torso ratio predisposes neonates to acceleration-deceleration injuries, more diffuse brain and upper cervical spine injuries, acute subdural hematomas, and air embolism [20]. The anaesthesiologist plays a vital role beginning from anaesthesia for diagnostic imaging such as CT scan, MRI, PET scan, preoperative assessment and optimization, and intraoperative management and the role extends further into postoperative neurocritical care.

40.4 Neuroembryology and Pathogenesis of Diseases of the CNS

The knowledge of embryology of the neural tube in fetal life is the foundation for understanding the genesis, clinical features, need for urgent neonatal surgical correction and the challenges encountered by the anaesthesiologist in the perioperative management of one of the most commonly encountered groups of neurosurgical conditions which comprise of **Neural Tube Defects, Arnold Chiari Malformations, and Hydrocephalus.**

40.4.1 Embryology and Pathogenesis of Neural Tube Defects (NTDs)

The development of CNS begins in the third week of gestation, around day 13–14 of embryonic life. The ectodermal cells rostral to the primitive node give rise to the neural plate. Bending of neural plate to form a pair of neural folds starts by 17–18 days post-fertilization. As the folds grow, they approach each other towards the midline, enabling fusion to occur, leading to formation of the neural tube (NT). Fusion of neural folds is a discontinuous process which starts at multiple but discrete points, starting at the junction of future hind brain and cervical spinal cord at day 22, extending bidirectionally, cranial and caudal. Another point of fusion is initiated at the rostral end of forebrain, from where it extends cranially and meets the upward closure wave from the hindbrain. Areas between the points that are initially not fused are known as neuropores. As the fusion progresses, neuropores shrink and close completely, ultimately leading to the formation of a closed neural tube in the fourth week. Cranial closure of rostral neuropore occurs earlier, by day 24, whereas closure of the posterior neuropore gets completed later, at day 26. This phase of formation of a completely closed neural tube is called **primary neurulation**. The neural tube gives rise to future brain and spinal cord till its upper lumbar level. Incomplete closure of neuropores during primary neurulation results in open NTDs. This phase of primary neurulation is followed by **secondary neurulation** which forms the neural tube in sacral and coccygeal regions. The caudal eminence of mesenchymal cells in tail end of embryo reorganizes to form longitudinal cell condensations. The dorsal portions of these cells (neural precursors) undergo canalization to form the secondary neural tube which is completed by the sixth week of gestation and gives rise to spinal cord at lower lumbar levels and extends till S2. Hence, secondary neurulation does not have a closure component. Most NTDs in this phase result from indistinct separation of neural and mesodermal tissues and are closed NTDs. The prevalence of NTDs is 1:1000 births, with regional variations [21–24].

40.4.1.1 Types of Neural Tube Defects

Spinal dysraphism ("Raphe" is a Greek word meaning line of union of two contiguous bilaterally symmetric structures) is a general term used for defects arising from incomplete fusion of neural tube and vertebral arches and is often used interchangeably with **Spina bifida**. It can be classified into:

Classification I

(a) **Spina bifida Cystica**: meningocele (outpouching of meninges) or myelomeningocele (outpouching of spinal neural tissue and meninges).
(b) **Spina bifida Aperta**: myeloschisis (absent or ruptured cyst).
(c) **Spina bifida Occulta**: external cutaneous lesion with defective fusion of vertebral arches. Neural tissue and meninges stay within the spinal canal.

In this classification, it is not possible to associate each class of spina bifida consistently with the neurological deficit. For instance, in spina bifida cystica, the

neurological deficit may be either markedly present as in myelomeningocele, or limited to absent as in meningocele. The only class of spina bifida that is associated with CNS deficit is spina bifida Aperta. Because of the non-clarity of kind of neurological deficit in the above classification, a more useful classification was proposed. The above defects resulted from defective fusion of neural tube and were hence termed collectively as NTDs

Classification II

1. **Open neural tube defect:** These defects occur due to faulty closure of neural tube during the neurulation phase, leading to exposure of neural tube tissue. Chronic exposure of neural tissue to amniotic fluid in utero has toxic effect causing neuronal tissue degeneration, interrupted axonal connections, and neuronal cell death leading to loss of neurological function and disability. Therefore, fetal surgical attempts to cover the open neural tube lesion during fetal life have been encouraged, so as to arrest or prevent neurodegeneration when complete neural tube closure has failed. These defects are associated with CSF leakage and ACM.
   (a) **Myelomeningocele**: The dysplastic meninges along with the spinal cord protrude out through a defect in the posterior vertebral arches and extend beyond the spinal canal.
   (b) **Myeloschisis**: the neural placode gets plastered with the ventral part of spinal canal. There is no cystic formation.
   (c) **Hemi myelomeningocele**: Split cord malformation with part of the cord closed, the other part is open causing protrusion of neural elements along with meninges. Defects in pre-neurulation as well as neurulation lead to this anomaly.
   (d) **Craniorachischisis**: Severe open NTD in which there is failure of initiation of closure of the neural tube at day 22.
   (e) **Anencephaly**: Failed closure of cranial rostral neuropore.
2. **Closed neural tube defect:** The neural tube is closed. Hence, it is not associated with CSF leakage or marked CNS defects or ACM.

Defects in Secondary Neurulation Phase (a)

1. **Posterior lumbar, sacral, and thoracic meningocele**: Abnormality in spine leads to outpouching of meninges. Occasionally, there can be neural elements such as aberrant nerve roots, ganglion cells, or glial tissue adhered to the inner surface of the dome of meningocele.
2. **Anterior sacral meningocele**: There is anterior herniation of the dura mater beyond the spinal canal leading to the development of the anterior meningocele. These are occult lesions without visible abnormalities. The cysts are small to start with, and slowly enlarge over decades, displacing the rectum, bladder, and ureters, giving rise to difficulty with bladder and bowel function and dystocia in females.
3. **Lipomatous malformation:** Most common form of closed NTDs charecterized by presence of excessive lipomatous tissue.
4. **Abnormal filum terminale:** Presence of an abnormal filum that is shorter, thicker and contains increased amount of connective and adipose tissues. It often

co-exists with lipomatous malformations and presents as Conus Medullaris syndrome.
5. **Congenital dermal sinus:** Occurs due to abnormal separation of neural ectoderm and cutaneous ectoderm. It is a tract lined by stratified squamous epithelium extending below the skin to subcutaneous tissue or deeper and often coexists with other cutaneous markers.
6. **Association with caudal regression:** Partial to complete absence of coccygeal, sacral, lumbar and lower thoracic vertebrae.

Defects in Primary Neurulation Phase (b)
1. **Posterior cervical meningocele and limited dorsal myeloschisis**: An abnormal midline endo mesenchymal tract grows that bisects the notochord and neural plate during the primary neurulation phase. Hence, there is a stalk with or without dysplastic glial elements extending to the dome or side of the meningocele sac.
2. **Myelo-cystocele**: This is an extension or a diverticulum from the central canal of spinal cord containing CSF that protrudes through a defect in the posterior vertebral arches to extend beyond the spinal canal; however, the spinal cord remains within the spinal canal. Common sites are posterior cervical and lumbosacral.
3. **Neureneteric (NE) cyst**: or enterogenous endodermal cysts, arising from more extensive disruption of the notochord and neural plate. However, they are rare, benign, with no neurological deficit.
4. **Split cord malformation**: Presence of two hemicords, each having a single set of laterally located dorsal and ventral nerve roots contained within two distinct dural sheaths [25].

40.4.2 Embryology and Pathogenesis of Arnold Chiari Malformation (ACM)

During the development of CNS, there is a temporary phase of spinal central canal occlusion for 2–8 days that occurs just prior to NT closure, hence CSF stays in the cranial compartment, under pressure, leading to distension of developing ventricles. This distension provides inductive stimulus for growth of cranial neuroectoderm and surrounding mesenchyme. End of this occlusion phase is preceded by complete fusion of the NT. Therefore, the required distending pressure for development of cranial structures, after the occlusion phase is over, is maintained by closure of NT once the occlusion reverses. Failed closure of NT causes failure to sustain this adequate distending pressure leading to miscoding of volume determination and consequently a small posterior fossa, impaired supratentorial neuronal development with malformed cerebrum, and dysgenesis of corpus callosum as well as mesenchymal defects. The ventricular fluid also drains via the open NT, leading to defective development of ventricles, consequent ventricular atresia and hypoplasia. Since the posterior fossa is not well-developed and is small, it is unable to accommodate the

developing cerebellum and brainstem. The hindbrain herniates cranially through the incisura compressing the aqueduct and breaking the collicular plate. Caudal displacement of the hind brain occurs through the foramen magnum into the cervical canal. The impaired development of ventricles along with impaired flow of CSF due to kinked aqueduct contributes to development of hydrocephalus. ACM is thus a precursor of hydrocephalus and 80% cases of ACM develop hydrocephalus, which can further exaggerate symptoms of hind brain compression. Overt hydrocephalus due to aqueductal stenosis is present in 15% neonates with MMC at birth. Closure of MMC defect will stop CSF drainage, thereby leading to progressive ventriculomegaly and precipitate hydrocephalus. The clinical manifestations of ACM are of hindbrain compression, brainstem dysgenesis, cranial nerve nuclei hypoplasia, and basal pontine nuclear hypoplasia, exaggerated by hydrocephalus. Presence of manifestations such as stridor, cyanosis, high pitched cry, abnormal breathing patterns, and feeding problems (nasal feed aspiration, choking while feeding, gastroesophageal reflux, and aspiration pneumonitis) are linked to poor outcomes.

The management of symptoms related to ACM II, central apnea, stridor, and vocal cord palsy involves CSF diversion as the first step and then consider decompression, if the neonate is normal at birth but develops acute symptoms despite effective CSF diversion [26, 27]. Closure of NTD promotes upward movement of herniated hindbrain. Treatment of hydrocephalus with shunting is crucial to prevent and reduce symptoms of hindbrain compression before undertaking decompression procedures. Anatomical malformations can lead to isolated lateral ventricles, leading to unilateral functioning of the shunt. Despite surgery, complete resolution of symptoms may not be possible since certain causative factors such as brain stem dysgenesis are not reversible and only hind brain herniation is amenable to surgical correction [28–32].

40.4.3 Embryology and Pathogenesis of Congenital Hydrocephalus

CSF maintains the internal environment of the brain and shields the brain from homeostatic disturbances, such as acute concentrations of serum electrolytes. CSF is produced by choroid plexus and secreted into the lateral ventricles at a rate of 0.3–0.4 mL/min across all age groups, except for being slightly lower in preterm neonates. Since the ventricular size is smaller in children than adults, turnover of CSF is also faster, and presence of non-communicating hydrocephalus leads to an acute rise in ICP in them. From lateral ventricles, CSF flows from the foramen of Monroe into the third ventricle, and then through the aqueduct of Sylvius into the fourth ventricle, from where CSF flows out via foramen of Mangendie medially and foramen of Luschka laterally, into the basal cisterns [33, 34].

Congenital hydrocephalus At can result from open NTD, Dandy-Walker syndrome, X-linked hydrocephalus, mucopolysaccharidosis, in utero IVH), and Maroteaux-Lamy syndrome. **Acquired hydrocephalus** can result from IVH, trauma, space occupying lesions (SOL) and infections. Preterm and LBW

neonates have high incidence (30–50%) of IVH that causes post-hemorrhagic ventricular dilation (PHVD) and Progressive post-hemorrhagic hydrocephalus (PHH) [35, 36].

Management options include, medical treatment with diuretics (most commonly used), and surgical or endoscopic shunt placement. Invasive options for hydrocephalus include:

(a) **Surgical Ventriculostomy or Ventricular shunt: Ventriculo-peritoneal (VP) or Ventriculo-atrial (VA) shunt**.
(b) **Endoscopic third Ventriculostomy (ETV)**.
(c) **External ventricular drain (EVD)**.
(d) **Neuro endoscopic ventricular lavage** [37, 38].
(e) **Ventriculo-subgaleal shunts**.

All these procedures are conventionally performed in the operation theatre (OT) under general anaesthesia (GA). **A Ventricular access device** placement can be done outside the OT, by the bedside, under local anesthesia (LA), at low risk, especially in preterm neonates, as it avoids the need of GA and intubation [39].

40.5 Preoperative Assessment as per the Neurosurgical Condition

40.5.1 Hydrocephalus

Assessment begins with detailed history from parents, which reveals excessive increase in head circumference, anterior fontanelle fullness especially in upright position, episodic apnea, bradycardia, general lethargy, and abnormalities of ocular movement (restricted up gaze). Symptoms suggestive of **chronic hydrocephalus** are increasing head size, irritability, poor feeding, vomiting, altered behaviour, and developmental delays. These symptoms are non-specific and combination of vomiting and decreased feeding can mimic gastroenteritis. The rise in head circumference can be severe enough to increase difficulty in handling the airway. The normal rise in head circumference in first month of life is 2 cm [40]. **Acute hydrocephalus** can manifest as vomiting, dehydration, sluggishness, and altered sensorium, and if left untreated, may result in cerebral and brainstem herniation, cardiorespiratory arrest, and even death. The commonest form of hydrocephalus seen in neonates is due to abnormal resorption of CSF, mostly associated with IVH. VP shunting should be done judiciously in hemorrhagic hydrocephalus due to its several disadvantages and complications. Non-hemorrhagic hydrocephalus due to aqueductal stenosis, posterior fossa cysts, holoprosencephaly, or hydranencephaly can be shunted within 1–2 days of birth, to prevent subsequent massive head enlargement.

40.5.2 Intracranial Cystic Spaces

Interhemispheric, temporal fossa, posterior fossa, and other arachnoid cysts may be found incidentally or may present with macrocephaly or hydrocephalus in neonates. Those causing symptomatic mass effect or accelerated head growth may be fenestrated, endoscopically or by open microsurgical technique, into the basal cisterns or ventricle. fourth ventricular cysts and placement of shunts to drain them can elicit dangerous brainstem responses like apnea and bradycardia.

40.5.3 Neural Tube Defects

Abnormal developmental folding of the NT and anterior neuropore can vary from benign (spina bifida occulta with no neurologic sequelae) to the most severe presentation (anencephaly, craniospinal rachischisis, complete absence of NT closure). Open defects, such as MMC, or defects that leak CSF or interfere with airway patency (large nasofrontal encephalocele, open occipital encephaloceles, those with impending rupture) require urgent repair within a few days of birth. Surgeries for tethered cord syndrome, spinal lipomas, and other closed malformations are typically delayed until the patient is at least 3 months age [41, 42].

Neonates with NTDs present with swelling anywhere along the spine region. History of fever and discharge indicates rupture or infection of MMC. Assessment of motor function can be done by asking if the movement of all limbs is equal or not. Lower cranial nerve dysfunction, indication of ACM, can be assessed by eliciting history of weak or poor cry, inability to swallow feeds and nasal regurgitation of feeds, stridor, and cyanosis (cyanosis could also indicate co-existent cardiac anomaly). MMC may be associated with other systemic anomalies. (Table 40.1) [43–47].

The location of MMC is relevant for predicting difficult mask ventilation (frontonasal encephalocele), difficult laryngoscopy and intubation (occipital encephalocele), and risk of postoperative respiratory palsy (cervicothoracic MMC). Neurological deficit is expected to be present below the level of MMC, with interruption of the spinal cord at the site of the MMC, and paralysis of the legs, incontinence of urine and feaces, anaesthesia of the skin, and abnormalities of the hips, knees, and feet [24]. Only 1% of newborns with open NTD are free of handicap. Incidence of seizures is 15–25% and is most likely related to cerebral anomalies (cortical heterotopias, polymicrogyria), which are associated with ACM II [28].

History should include birth history (gestational age, birth weight, APGAR), exposure to risk factors for occurrence of NTD (antenatal exposure to drugs (anticonvulsants) or radiation, folic acid supplementation, similar defects in sibling / family members), maternal diabetes, and obesity [23]. Milestones achieved in the first month of life should be documented. Lastly, it is important to take history of vaccinations received as per the national guidelines. A detailed **general and systemic examination** of neonate should be done. Ascertain the size and location of MMC to predict the magnitude of intraoperative fluid shifts and blood loss. MMC

Table 40.1 Anomalies associated with MMC

Trisomy 18	**Common (>75%):** Cardiac (septal defects, PDA, poly valvular disease) **Frequent (25–75%):** Genitourinary (horse shoe kidney) **Less frequent (< 25%):** Gastrointestinal (omphalocele, TEF, pyloric stenosis, Meckel's diverticulum), CNS (cerebellar hypoplasia, corpus callosum agenesis, spina bifida, polymicrogyria), craniofacial (orofacial clefts), ocular (microphthalmia, coloboma, corneal opacities, cataract), limb (radial aplasia/ hypoplasia)
Trisomy 13	**Face**: Sloping forehead, small malformed ears, anophthalmia or microphthalmia, micrognathia, pre-auricular tags **CNS**: Midline anomalies, alobar holoprosencephaly **Limbs**: Postaxial polydactyly, congenital talipes equinovarus **CVS**: Septal defects, tetralogy of Fallot (TOF), double outlet right ventricle **Others**: Cryptorchidism, hypospadias, labia minora hypoplasia, bicornuate uterus, omphalocele, incomplete rotation of the colon, Meckel's diverticulum, polycystic kidney, hydronephrosis, horseshoe kidney
Meckel Gruber syndrome	Occipital encephalocele, bilateral large multicystic kidneys, fibrotic changes of liver, Polydactyly
HARD syndrome	Hydrocephalus, Agyria, retinal dysplasia
VACTERL group	Vertebral defect, anal atresia, cardiac defect, TracheoEsophageal fistula, renal anomalies, and limb abnormalities
OEIS complex	Omphalocele, Exstrophy, imperforate anus, and spinal anomalies
Jarcho-Levin syndrome	NTD, vertebral body/rib malformation, kyphosis, scoliosis, short stature short necks, limited neck movement, difficulty breathing, small malformed chest (crab-like appearance), defects – CNS, cardiac, genitalia, syndactyly, camptodactyly, typical facial features
Roberts (pseudothalidomide) Syndrome	Malformation of the bones in the skull, face, arms, and legs – Tetraphocomelia, growth retardation, hypoplasia, oligo -syn -clinodactyly, cleft lip-palate, elbow-knee contractures, micrognathia, exophthalmos, encephalocele, ear malformations, intellectual disability

and meningoceles are usually pedunculated and have positive transillumination test. Ruptured MMC with fever and pus discharge makes the neonate more susceptible to intraoperative hemodynamic disturbances. Raised ICP features may be present. CNS evaluation includes level of consciousness, measurement of head circumference, and cranial nerve functions. CVS examination is done to rule out cardiac defects. Look for abnormal facial features (may further increase airway difficulty), polydactyly, and gross anomalies of extremities. **Preoperative investigations** should include haematological (Hb to calculate maximal allowable blood loss and total leukocyte count to rule out infection), Biochemical (blood sugar, liver enzymes, serum bilirubin, blood urea, serum creatinine, electrolytes, calcium), Blood grouping and crossmatching, and others (USG abdomen, ABG, Echo) [48], radiological

(X-ray head and chest, CT, MRI, PET scans of brain and spine), and Transcranial doppler (to predict cerebral perfusion in babies with raised ICP). Altered flow signals in anterior cerebral artery upon compression of anterior fontanelle, at transcranial doppler study, suggest changes in ICP warranting shunt placement [49].

40.5.4 Craniosynostosis

Premature intrauterine fusion of one or more cranial sutures with abnormal skull growth causes craniosynostosis. Surgical management includes **strip (open) craniectomy** which should be carried out at the age of 3–6 months. Anaesthetic concerns include difficult airway, sudden and massive blood loss, and venous air embolism (VAE) during surgery, and facial oedema, need for sedation and pain management, total parentral nutrition or Ryles tube feeds, and ventilatory support in the postoperative period.

Multiple suture craniosynostosis may be associated with craniofacial abnormalities such as Apert's, Crouzen's, and Pfeiffer syndromes. Surgical correction of the syndromic categories is usually deferred for several months beyond the neonatal period. The **kleeblattschädel syndrome** (cloverleaf skull), when all sutures are prematurely closed, is the only craniosynostotic syndrome requiring treatment in the neonatal period. Currently, endoscopic procedures for single suture involvement are being carried out to prevent morbidities of open suturectomy. **Endoscopic strip craniectomy** is minimally invasive, used recently for surgical correction of craniosynostosis that offers advantages of decreased magnitude of blood loss and improved wound healing. This procedure has been carried out successfully in infants with age as low as 4–5 weeks [50, 51]. The advancement in minimally invasive surgical techniques, monitoring techniques, and neurocritical care, in conjunction with a multidisciplinary approach comprising of neonatologist, pediatric neurosurgeon, and pediatric neuroanaesthesiologist, can pave way for correction of craniosynostosis in the neonatal period in the coming future.

40.5.5 Large Cerebral Arteriovenous Shunts

The most frequent cerebrovascular anomaly in neonates is the **vein of Galen aneurysmal malformation (VGAM)**. Arteriovenous malformations (AVMs), AV fistulas (Dural and pial), and cavernous malformations have also been described. In neonates, VGAM often presents with high output cardiac failure requiring inotropes, pulmonary hypertension, and myocardial ischemia. Initial treatment of a high-flow fistula involves **endovascular embolization** by trans arterial or transvenous route in the radiology suite. Surgical resection of these vascular lesions is associated with massive blood loss, sudden postoperative hypertension, and hyperemic cerebral oedema, necessitating vasodilator therapy for the hypertensive crisis [52, 53].

40.6 Anesthetic Concerns in a Neonate Undergoing Neurosurgery

Neonates have an increased perioperative risk due to;

1. Emergency conditions of surgery.
2. Presence of undiagnosed congenital anomalies.
3. Persistence of transitional circulation in premature neonates.
4. Intracardiac shunting through patent ductus arteriosus or unclosed foramen ovale.
5. Possible congestive heart failure (CHF) associated with large cerebral arteriovenous (AV) malformations.
6. Airway difficulty - Management of neonatal respiratory system may be difficult because of the small-sized airway, craniofacial anomalies, laryngotracheal lesions, acute respiratory disease due to hyaline membrane disease or retained amniotic fluid, or chronic respiratory disease like bronchopulmonary dysplasia.
7. The immature neonatal organ systems, especially the myocardium which is highly sensitive to anaesthetic drugs and surgical stress.
8. Not-fully developed hepatic and renal systems.

Craniotomy for total or partial resection of brain tumours is a major, prolonged surgery with risk of hypothermia, hypoglycemia, fluid shifts, and massive blood loss. Neonatal brain tumours are usually infratentorial that mandate use of prone position. Poor intracranial compliance necessitates reduction in ICP and a relaxed brain that can be achieved by maintaining adequate depth of anaesthesia, hyperventilation and moderate hypocarbia, use of diuretics (Mannitol (0.5–2 g/kg), and/or Furosemide (0.5 mg/kg)), and head elevation. Preoperative embolization of choroid plexus tumours also helps reduce intraoperative blood loss and protect the vulnerable neonate from hemodynamic instability [18, 19].

The anaesthetic management for neurosurgery in neonates is impacted by:

(a) **Patient Factors** - Concerns related to the neonatal age, unique multi-system anatomical and physiologic variations, particularly of the CNS, and possible co-existing anomalies,
(b) **Disease Factors** - Concerns due to the underlying neurosurgical disease and its pathology, and.
(c) **Surgical Factors** - Concerns imposed by the neurosurgical procedure itself, craniotomy and open surgery, or endoscopic procedure.

40.6.1 Anesthetic Concerns Due to Neonatal Age

The neonate is not merely a miniature adult. There are several anatomical and physiological differences in all systems of the body when compared with adult which can have several anaesthetic implications. Although these have been discussed in

detail in previous chapters, these general concerns have many unique implications when a neonate has to undergo a neurosurgery. Specific implications pertinent to neurosurgery are listed in Box 40.1.

Box 40.1 List Specific Implications Pertinent to Neurosurgery in a Neonate

- Presence of craniofacial abnormalities.
- Difficult airway.
- Prematurity and low birth weight – Immature CNS, liver, kidney, etc., postop apnoea.
- Associated congenital heart disease - hypoxemia, paradoxical air embolism, arterial air embolism.
- Hemodynamic disturbances - arrhythmias, hypotension, venous air emboli.
- Gastrointestinal reflux - aspiration pneumonia.
- Upper respiratory tract infection causing laryngospasm, bronchospasm, hypoxia, pneumonia.

40.6.1.1 Airway and Respiratory System

(a) Difficult airway: The airway in neonates is already an anticipated difficult airway, difficult mask ventilation, difficult laryngoscopy, and difficult intubation. Presence of hydrocephalus, occipital encephaloceles, and cervical MMC may further impede neck movements. Frontonasal encephaloceles result in difficult mask holding and difficult mask ventilation. Presence of syndromes with abnormal facial features can also impair airway handling. Hence, the already difficult airway of neonate can become even more difficult to handle. The difficult airway, coupled with a combination of low FRC (25 mL/kg, adults 35 mL/kg) and high metabolic O_2 demand, results in reduction of the safe apnea time, defined as time duration between last breath until SpO_2 falls below 90%, and consequently early desaturation. Safe apnea time of 25 s is reported in preterm neonates [54]. Thus, the importance of a meticulously performed preoxygenation with 100% O_2, which can prolong safe apnea time, cannot be overemphasized. However, attaining effective preoxygenation ($FeO_2 > 0.9$) itself is challenging because of non-compliant age group. Judicious use of sedation titrated so as to just facilitate preoxygenation can be tried to achieve target FeO_2.

(b) Due to higher alveolar ventilation (VA) to FRC ratio (**VA:FRC**), preoxygenation occurs faster in neonate; however, total O_2 reserves created are lower. Therefore, despite achieving target FeO_2, neonates remain at risk of **early desaturation**. Employment of apneic oxygenation methods from an auxiliary O_2 source during the entire apneic period (while laryngoscopy and intubation are being performed) can further prolong the safe apnea time.

(c) Apart from the risk of early desaturation, airway handling especially laryngoscopy and intubation can result in **hemodynamic instability** [55]. Laryngoscopy and intubation performed by an experienced anaesthesiologist trained in neonatal intubations and use of muscle relaxation can reduce the incidence of intubation-related adverse events [56].

(d) Impairment of lower cranial nerves in neonates with ACM and slow gastric emptying in presence of raised ICP can potentially increase the **risk of aspiration** in unprotected airway.

(e) Lungs are immature with **reduced compliance** and **increased work of breathing**. Mainstay of ventilatory strategy is thus use of lower tidal volumes, lower inspiratory pressures, use of low PEEP to avoid atelectasis, and higher respiratory rates, with acceptance for mild hypercapnia ($PaCO_2$ 45–55 mmHg) [57]. Application of **PEEP** and use of hypercapnia may not be tolerated by neonates with raised ICP. It is recommended to avoid hypocapnia ($PaCO_2 < 39$ mmHg), hypercapnia ($PaCO_2 > 60$ mmHg), as well as sudden fluctuations in $PaCO_2$ values during first 4 days of after birth, especially in preterm neonates who are more prone to develop IVH. Oxygen must be used judiciously because excessive O_2 predisposes to injury of the developing brain, although there are no conclusive outcome studies demonstrating poor neurodevelopmental outcomes in neonates receiving liberal O_2 (target SpO_2 91–95%) compared with neonates receiving restricted O_2 (target SpO_2 85–89%). There are no definite guidelines regarding the target SpO_2 for neonates. Most acceptable safe limits range from 87–88% up to 94%.

(f) **Length of trachea** is short, around 5 cm. Shorter tracheal length narrows the margin of safety for proper ETT positioning and predisposes both to endobronchial intubation with neck flexion and accidental extubation with neck extension. Either of these manoeuvres is usually required to obtain ideal surgical position during neurosurgery. Thus, presence of bilateral equal air entry by chest auscultation must be reconfirmed after final head positioning.

(g) The central control of respiratory rhythm and central and peripheral chemoreceptors are immature making the neonate susceptible to **postoperative apnea**. Preterm neonates experience apneic episodes up to 60 weeks postconceptional age. Breathing hypoxic mixture leads to a short period of hyperpnea followed by prolonged bradypnea and apnea. Ventilatory response to hypercapnia is present in term and is weak in preterm neonates.

(h) These factors along with brainstem pathologies and depressant effects of anaesthesia on respiratory system make the preterm and term neonate even more susceptible to **apnea and respiratory depression in the postoperative period**.

(i) **Delayed tracheal extubation** is expected in ACM and brainstem surgery due to intermittent postoperative apnea, vocal cord paralysis, respiratory irregularities, significant airway oedema, and pre-existing bronchopulmonary dysplasia [58].

40.6.1.2 Cardiovascular System (CVS)

(a) **Hemodynamic changes in neonates:** Cardiac output, which is a product of stroke volume and heart rate, is rate-dependent in neonates, therefore maintaining normal sinus rate and rhythm is crucial to maintain cardiac output and thus CPP. MAP and SBP increase by 8 mmHg in first 72 h of life in term as well as preterm infants. Also, hypotension even in non-anaesthetized neonates is not well-defined. **Physiological hypotension** is defined as MAP below which cere-

brovascular autoregulation is lost leading to neonatal cerebral ischemia. Most widely used definition of hypotension is MAP below 5–10 percentile of MAP at a particular gestational or postnatal age. MAP above 30 mmHg is maintained in premature (born at 26–30 weeks) neonates to reduce occurrence of IVH. A rough guide to target MAP is the gestational age in weeks, and goal is to maintain MAP above the target value. Brainstem manipulations are accompanied by severe cardiac manifestations in the form of bradycardia, arrythmias, and sudden cardiac arrests.

(b) Neurosurgery can be associated with **major fluid shifts**. Estimation of fluid loss and adequate replacement of losses are important since baroreceptor responses to circulating blood volume are immature. Thus, hypovolemia does not lead to compensatory tachycardia to maintain cardiac output, and hypotension ensues with decrease in CPP. Over-correction with fluids can predispose to congestive heart failure (CHF) since left ventricle (LV) is less compliant and cannot tolerate overload.

(c) Hemodynamic monitoring concerns: Measurement of BP in neonate is technically challenging. The value of NIBP will be affected by the limb used and type of instrument. The bladder cuff width in oscillometer NIBP measurement technique should be half of mid arm circumference. 20 mmHg lower MAP in legs (than arms) in 8% of neonates and different readings between the two upper arms in 16% neonates have been reported with non-invasive methods [59, 60]. The gold standard method is MAP measurement, using invasive intra-arterial catheter. It is recommended in all critically ill neonates and if MAP recordings with oscillometer are less than 30 mmHg [61]. Hence, in neonates with neurosurgeries where major fluid shifts are expected such as large MMC, use of IABP in place of NIBP would be useful. For anaesthetized neonates, hypotension is defined as MAP less than 20% of baseline. Absolute target range for minimum SBP in neonates is 45.5–49.6 mmHg [62, 63].

(d) Perioperative hypoxia, hypercarbia, acidosis, hypothermia, and hypoglycemia can lead to **transition to fetal circulation** and **pulmonary hypertension**, which worsen perioperative outcome.

40.6.1.3 Hepatorenal System

Liver and Kidney are immature, and mature by the age of 2 years. GFR in term neonates is 35% of adults, with prolongation in drug clearance times of most drugs with prolonged duration of action. This can interfere with recovery and on-table extubation that is important after neurosurgery to assess the neurological outcome by a focussed post-extubation neurological evaluation of the neonate.

40.6.1.4 Thermoregulation

High surface area to volume ratio, less subcutaneous fat, lack of shivering, all make the neonate vulnerable to **hypothermia** especially during prolonged surgeries, and most neurosurgical procedures fall in this category. Hypothermia further delays anesthetic drug elimination, delays recovery, and interferes with early extubation post-neurosurgery. It also increases blood viscosity and shifts

ODC (oxygen dissociation curve) to left, with impaired cerebral O_2 delivery, transition to fetal circulation, and potential to increase ICP further. Opening up of functionally closed intracardiac shunts also predisposes them to systemic arterial embolism after paradoxical venous air embolism, cerebral embolism, and cerebral hypoxemia. All measures to prevent hypothermia are integral to neuroanaesthesia.

40.6.2 Anesthetic Concerns Due to Neurological Pathology

The neurological lesions induce multi-system abnormalities that need to be addressed in the perioperative period, e.g. lower cranial nerve involvement (altered sensorium, aspiration pneumonitis), brainstem lesions (respiratory insufficiency, cardiovascular abnormalities), pituitary lesions (diabetes insipidus, hypothyroidism, adrenal insufficiency), ACM (stridor, apnea), AV malformations (congestive cardiac failure), excessive vomiting (electrolyte imbalance), and seizures.

1. **Maintaining cerebral blood flow (CBF):** CBF is determined by CPP and cerebrovascular resistance (CVR). Preventing rise in ICP is crucial for maintaining CPP. In absence of an IV access, inhalational induction can be done to avoid rise in ICP due to crying of neonate at IV-line placement. Inhalational induction, laryngoscopy, intubation, and incision are times when increase in ICP can occur. Drugs and anaesthetic techniques can be deployed to maintain adequate plane of anaesthesia and prevent rise in ICP at these time points. Increase airway pressures (intraoperative bronchospasm) and application of PEEP increase intrathoracic pressures, thereby increasing CVP and decreasing CPP. Hence, maintaining target MAP at all times, during induction and in the intraoperative period, is essential to maintain CPP.
2. **Brainstem dysfunction**- Neonates with ACM have brainstem dysgenesis and dysfunction that predispose to perioperative hemodynamic instability and postoperative stridor and apneic episodes.
3. **Implications of drugs** - Antiepileptics can induce hepatic enzymes and increase the requirements of anaesthetic drugs, diuretics such as acetazolamide, lasix and mannitol can lead to dyselectrolytemia, and steroids can be associated with metabolic disturbances.
4. **Maintaining cerebral O_2 delivery** to prevent cerebral hypoxia - $CMRO_2$ or cerebral O_2 consumption can be reduced by maintaining adequate depth of anaesthesia, and use of antiepileptics, if indicated. Cerebral O_2 delivery can be ensured by maintaining O_2 flux and cerebral blood flow. Hb concentration, PaO_2, and cardiac output are the major global determinants of cerebral blood flow and O_2 concentration. Prompt replacement of lost blood is needed.
5. **Cerebral autoregulation** is impaired by inhalational agents, hypercapnia, and vasodilators [64].

40.6.3 Anesthetic Concerns Due to Neurosurgical Intervention

Neurosurgery in the neonate generates several perioperative concerns depending on the approach used, craniotomy vs endoscopic, from prolonged duration of surgery, sharing of the airway field, intraoperative position-related, excessive blood loss, haemodynamic disturbances, raised ICP, venous air embolism (VAE), hypothermia, delayed awakening, and seizures due to pneumocephalus. The most commonly encountered conditions requiring neurosurgical treatment in a neonate are excision of MMC, decompression of ACM-II, and shunt insertion of congenital hydrocephalus. Early postnatal repair of MMC should ideally be performed within first 48 h of neonatal life to minimize further neurological damage, risk of infection, and rupture of MMC [23, 24, 65, 66].

ACM is characterized by caudal displacement of cerebellum, fourth ventricle, and medulla into the cervical spine, and it co-exists in almost all neonates (>90%) with MMC; however, clinical manifestations are present in 15–35% neonates with MMC. Presence of symptoms of MMC, especially stridor, worsens the prognosis even after decompression surgery. Kyphosis is the commonest spine anomaly associated with MMC and kyphectomy may need to be performed simultaneously with MMC repair in the neonate [67]. The common concerns during neurosurgery are as follows:

1. Neurosurgeries are usually procedures with **long time duration** with consequent risk of perioperative hypothermia, blood loss, and drug accumulation; all contributing to delayed awakening after surgery.
2. There is an unavoidable **sharing of the airway field** with the surgeon with ETT malpositioning and kinking, and accidental extubation due to manipulation of the head during surgery. Proper communication between neurosurgeon and neuroanaesthesiologist before head movements, gentle head movements when required, and close monitoring of respiratory parameters ($EtCO_2$, airway pressures, ventilator waveforms) aid in prevention and early detection of these mishaps. Special ETT such as flexometallic and south pole RAE tubes can be considered as these can be directed away from patient's head and operative field and are kink-resistant. Use of cuffed ETT has several advantages such as better optimization of ventilation, accurate $EtCO_2$ monitoring, minimal air leak, continuous lung recruitment, and prevention of aspiration; however, their use is associated with development of subglottic stenosis in 0.3–11% neonatal population. Special cuffed ETT with low-pressure, high-volume cuffs (microcuff tubes) have reduced the incidence of cuff pressure-related complications and subglottic stenosis.
3. **Blood loss**: Except for ventriculoperitoneal (VP) shunt insertion, most are major procedures, associated with significant blood loss, hypovolemia, decreased cardiac output, and CPP. Also, reduction in Hb leads to fall in O_2 content of blood, decreasing cerebral O_2 delivery. Blood grouping and cross matching are a must during preoperative preparation.

4. **Surgical positioning:** This is a huge problem in neonates due to their diminutive size, increased risk of fractures, and epidural hemorrhage due to thin skull. **Prone position** is deployed for operating on posterior fossa surgery and NTD. It increases the risk of accidental extubation, ETT kinking, pressure neuropathies, and ocular sequalae. Therefore, proper eye padding, securing of ETT with its connectors, and padding of various pressure points are prerequisites. Careful placement of proper sized bolsters should be done to avoid abdominal compression that can raise the epidural venous pressures and increase in surgical site bleeding. Inadvertent pressure on the chest from a large bolster can impede ventilation and should be avoided. Neurosurgeries in head region often require change in head positions from flexion to extension and lateral rotation. In adults, neck flexion is permitted till the point where at least two fingers can be insinuated between the chin and chest, but there is no such definite end point for neonatal population. Excessive flexion can cause endobronchial intubation, ETT kinking, and obstruct cranial venous drainage leading to decrease in CPP. **Lateral rotation** of neck is permissible up to 45° in adults. Further rotation can cause obstruction to ipsilateral vertebral vessels. Excessive flexion and extension can lead to brainstem compression and aggravate hemodynamic instability. VP shunt insertion is performed in **supine position** with neck extension. Care should be taken to avoid accidental extubation.
5. **Haemodynamic disturbances** like arrythmias or sudden cardiac arrests.
6. **Raised intracranial pressure during surgery**.
7. **Venous air embolism** (VAE); more likely in surgeries using head elevated position.
8. The neonate is a **small individual** who can get hidden under surgical drapes. It is imperative that the anaesthesiologist should have unrestricted access to ETT, ventilatory circuits, and IV lines. Before handing over the neonate to the surgeon, check that all connections are tight and well-secured.
9. **Hypothermia**,
10. **Delayed awakening**,
11. Seizures due to **pneumocephalus**.

40.7 Effects of Anaesthetic Agents on Cerebral Perfusion

40.7.1 Intravenous Agents

Barbiturates decrease CBF by decreasing $CMRO_2$ and direct cerebral vasoconstriction, decreasing cerebral blood volume (CBV), and thus decreasing ICP in accordance with Monroe Kelly Doctrine. Caution must be exercised to avoid fall in CPP secondary to fall in MAP. Autoregulation and cerebrovascular reactivity to CO_2 are maintained.

Thiopentone has neuroprotective properties by virtue of attenuation of ischemia-induced glutamate release, inhibition of cortical intracellular calcium increase, and

free radical scavenging activity. **Propofol** also decreases $CMRO_2$, CBF, CBV, and thereby decreases ICP. Fall in SVR and cardiac output cause fall in MAP and consequent fall in CPP. Cerebral autoregulation and cerebrovascular reactivity are preserved. Neonates are vulnerable to propofol-induced hypotension that may be severe and persistent and must be avoided [68].

Etomidate reduces $CMRO_2$, CBF, CBV, and ICP. It causes lesser cardiovascular instability than propofol and thiopentone, with no decrease in MAP and CPP. However, it causes severe cerebral vasoconstriction that can lead to cerebral ischemia and also causes adrenocortical suppression. These properties warrant cautious use of etomidate in neonates during neurosurgery.

Ketamine increases $CMRO_2$, CBF, CBV, and ICP and should be avoided in cases with raised ICP.

40.7.2 Volatile Anaesthetic Agents

Nitrous oxide- N_2O increases $CMRO_2$ and causes cerebral vasodilation, preferentially in the supratentorial grey matter and consequent increase in regional blood flow. This is mediated via sympathoadrenal stimulation and mitochondrial activation. Increased CBF can lead to rise in ICP. N_2O impairs cerebral autoregulation and can also impair CO_2 reactivity of cerebral vasculature, especially in conjunction with other volatile halogenated agents.

Halogenated agents: Like most of the anaesthetic agents, volatile anaesthetic agents (VAA) cause decrease in $CMRO_2$ that is expected to decrease CBF. However, these agents cause direct cerebral vasodilation and increase in cerebral perfusion, which in excess of metabolic O_2 needs is known as **luxury perfusion**. The magnitude of increased CBF by an inhalational agent is determined by the balance between direct vasodilation and potential of the agent to decrease $CMRO_2$. **Halothane** has greatest propensity to increase CBF. In children, this increase is known to persist even after halothane concentration has been reduced, leading to **cerebrovascular hysteresis. Isoflurane, desflurane,** and **sevoflurane** decrease $CMRO_2$ more than halothane and thus lead to lesser increase in CBF than halothane. However, children are more sensitive to the vasodilatory effects of volatile agents on brain. Cerebrovascular autoregulation is also impaired by volatile agents. Sevoflurane has the minimum increase in CBF, maintains cerebral autoregulation at lower concentrations (<1.0 MAC), and has better preservation of cerebral vasculature CO_2 reactivity and is therefore the preferred agent for inhalational induction. Sevoflurane and isoflurane are the preferred agents for maintenance of anaesthesia [33].

40.7.3 Opioids and Sedatives

Cause minimal changes in $CMRO_2$, CBF, CBV, and ICP. They blunt catecholamine release and rise in ICP following laryngoscopy and intubation. Premedication with opioids can cause respiratory depression and hypercapnia, leading to cerebral

vasodilation and rise in ICP. Hence, it should be used under supervision and monitoring. **Morphine** causes stimulation of Edinger Westphal nucleus and interferes with pupillary assessment. Pethidine metabolite, norpethidine, has proconvulsant action and is excreted via kidneys. Renal immaturity predisposes to accumulation of proconvulsant metabolite. **Fentanyl**, **remifentanil**, **alfentanil**, and **Sufentanyl** are the preferred opioids for their shorter duration of action. Remifentanil has a very short half-life and short context-sensitive half time. It can be given as an infusion and effect wears off within minutes of cessation. Fentanyl achieves peak brain concentration 4 min after its administration with effect lasting 15 min.

Benzodiazepines cause decrease in $CMRO_2$, CBF, CBV, and ICP. They are useful for sedation for diagnostic procedures and in babies on ventilator.

40.7.4 Muscle Relaxants

Succinylcholine raises ICP which can be offset by opioid premedication, additional incremental doses of thiopentone or propofol, and mild hyperventilation. It is useful in situations of difficult airway.

Non-depolarizing muscle relaxants that cause sympathetic stimulation can potentially lead to increase in ICP. Pancuronium increases the heart rate, Vecuronium causes bradycardia, while Rocuronium has minimal effect on heart rate and rhythm. All these drugs are metabolized in the liver and excreted by the kidneys. Atracurium and Cisatracurium do not rely on hepatorenal excretion and are preferred, as they will have minimal effect on recovery after surgery, allowing better neurological assessment of the neonate.

40.8 Anesthesia Technique: Basic Principles

40.8.1 Preoperative Fasting

Neonate should be kept fasting using 6-4-2 rule that means at least 6 h for formula milk, 4 h for breast milk, and 2 h for clear liquids [69]. Neonates with ACM and lower cranial nerve dysfunction are prone to aspiration. Gastric emptying can also be slowed by raised ICP. Such neonates may be considered as full stomach and candidates for rapid sequence induction, in the modified (mRSI) or controlled RSI (cRSI) form [70].

40.8.2 Premedication

Premedication is seldom administered in neonates; first, because of absence of separation anxiety, and second, unsupervised administration can cause sedation leading to respiratory depression, hypercapnia, and rise in ICP. Premedication just prior to induction of general anaesthesia under monitoring in the OR can be administered to blunt responses to laryngoscopy and intubation.

40.8.3 OT Preparation

Apart from routine check of OR, preparation for neonate undergoing neurosurgery includes control of ambient OT temperature, warming instruments and techniques, and readiness with age-appropriate difficult airway aids that includes appropriately sized cuffed and uncuffed endotracheal tubes, stylets, video laryngoscope with blades for neonatal age group, and flexible fibreoptic bronchoscope. Above all, presence of experienced anaesthesiologists and OT personnel cannot be undermined.

40.8.4 Induction of Anesthesia

The prime goal at induction of anaesthesia in neonates undergoing cranial surgeries is to maintain CPP and prevent increases in ICP, and these can be achieved by correct choice of drugs, avoidance of fall in MAP, maintaining eucapnia, avoidance of high pressure during bag mask ventilation (BMV), maintaining adequate depth of anaesthesia, and reflex suppression with adequate muscle relaxation and good analgesia.

40.8.5 Inhalational induction

Inhalational induction is preferred in neonates who do not have IV line in place. Neonates have high alveolar ventilation to FRC ratio, making inhalational induction faster, and also avoid ICP rise from neonatal crying. However, inhalational agents are known to cause increase in CBF secondary to cerebral vasodilatory effect, despite decrease in $CMRO_2$, and increase in ICP. Halothane has the highest potency to increase CBF, followed by isoflurane, desflurane, and lowest with sevoflurane. Hence, Sevoflurane is the preferred induction agent with its favourable cerebral effect profile and lower MAC.

Induction can be done with 1% sevoflurane to start with, and 1% increment every 4–6 breaths until neonate is induced, or alternatively, with 8% sevoflurane for rapid induction. Simultaneously, ECG, NIBP cuff, and pulse oximeter should be attached, along with establishing an IV access, preferably in lower limbs, for easy access. IV fentanyl (2–4μg/kg) along with muscle relaxant (vecuronium bromide or atracurium besylate) to facilitate tracheal intubation should be administered, followed by reduction in sevoflurane concentration. If cRSI is being opted for, rocuronium 1 mg/kg can be used to achieve intubating conditions at 1 min. Succinylcholine (1–2 mg/kg) can be used in neonates with anticipated difficult airway along with additional dose of IV thiopentone or propofol, and mild hyperventilation, to offset the rise in ICP.

Intravenous induction is preferred if an IV access is already in situ, beginning with fentanyl 2–4μg/kg, as premedicants, 5 min prior to induction, to achieve its peak effect, followed by thiopentone (5–7 mg/kg) or propofol (1–2 mg/kg). After confirming successful BMV, muscle relaxant of choice is administered.

40.8.6 Bag-mask Ventilation

Bag-mask ventilation precautions include:

- Avoiding pressure over the eyes and soft tissues of neck as this can impede venous drainage from head and increase CVP, reducing CPP.
  Hyperventilation can lead to hypocapnia and decreased CBF and must be avoided.

40.8.7 Laryngoscopy and Intubation

After ensuring adequate relaxation using a neuromuscular monitor if available, gentle and swift direct laryngoscopy and intubation should be performed by an experienced anaesthesiologist using an adequately sized cuffed ETT (micro cuff) and correct position confirmed by five-point auscultation before it is secured safely. ETT position is again confirmed after final positioning, before the baby is draped. Avoid placing an oral airway to secure the ETT, as it can lead to tongue edema and macroglossia by the end of surgery, which may cause airway obstruction after extubation. A soft bite block may be inserted if neonate is being placed in prone position.

40.8.8 Anesthesia Maintenance

Controlled ventilation using pressure control or volume control mode with O_2 air mixture and FiO_2 to target SpO_2 of 87–94% can be used. N_2O has the potential to raise ICP and worsens the effect of VAE, hence is avoided. However, there are no outcome studies negating the use of N_2O in neurosurgery. Spontaneous ventilation comes with the risk of raised ICP from coughing or bucking, increased chances of VAE due to generation of negative intrathoracic pressures during inspiration, decreased CPP and MAP as an effect of increase in inhalational anaesthetic drug requirement due to more myocardial depression in a neonate who is already sensitive to the cardio-depressant effects of volatile agents, and increase in ICP due to cerebral vasodilatation. Moreover, neonates generate auto-PEEP by various mechanisms when awake to maintain their FRC and prevent atelectasis, and this is abolished in spontaneously breathing neonates under general anaesthesia. Therefore, spontaneous ventilation is not recommended during neurosurgery.

Volatile agents for maintenance can be used for anaesthesia maintenance at concentrations up to 1.0 MAC that do not increase ICP. Isoflurane, desflurane, or sevoflurane can be used.

TIVA for maintenance leads to better control of ICP and CPP, especially in neonates where intracranial compliance is reduced. Propofol, fentanyl, and remifentanil are the agents for TIVA. Dexmedetomidine is approved by FDA for use in neonates and is neuroprotective. But there is not much evidence on its use as an agent for TIVA in neonates for long surgeries.

There is no evidence suggesting preference of TIVA over inhalational maintenance in neonates.

Neonate is then positioned for the surgical procedure, pressure points and eyes padded and protected, and ETT and circuit connections secured tightly, avoiding any drag on the ETT. Check that all monitoring equipment is well-placed and secured. Also confirm that IV, arterial, central venous lines, and urinary catheters are also well-secured and without kink or drag on them after positioning.

40.8.9 Prevention of Hypothermia

Prevention of hypothermia must be done using warmed mattress, fluid warmers, warm humidified breathing gases, warm fluids for surgical irrigation, body warmers, overhead radiant heaters, covering of all exposed parts of the neonate body, etc.

40.8.10 Analgesia

Analgesia can be provided with IV paracetamol, hourly supplements of short-acting opioids (fentanyl or remifentanil), local infiltration at the site prior to incision, and repeated at closure, depending on surgical duration. Epinephrine (1:200,000) when added to LA solution provides vasoconstriction, helps reduce bleeding, and prolongs its duration of action. Specific nerve blocks can be employed too; supraorbital and supratrochlear nerve blocks provide analgesia from frontal area to midcoronal portion of the occiput, and greater occipital nerve block provides analgesia from posterior of the occiput to the midcoronal area of the occiput, though multimodal analgesia is the modern pain management technique advocated and used. However, in neonates with low body weights and low gestational age, the safe dose limit of LA drugs should not be exceeded to prevent toxicity, remembering the potentiation of effect by other concomitantly administered analgesics, and that there should be no effect on recovery and postoperative neurological assessment.

40.8.11 Intraoperative Monitoring

Intraoperative monitoring must consist of non-invasive ECG, $EtCO_2$, SpO_2, temperature, NIBP, and urine output if indicated, and agent monitoring to keep MAC below 1.0. Monitoring of EtO2 should be done during preoxygenation and it is deemed adequate when EtO_2 of 85–90% is achieved. In event of VAE, EtN_2 falls earlier than $EtCO_2$. Hence, EtN_2 monitoring too is indicated for early detection of VAE. FiO_2 should be monitored throughout the surgery to avoid delivery of hypoxic mixtures and of very high FiO_2 for long periods because of the risk of ROP and BPD. In most cases, non-invasive monitoring suffices.

(a) **Invasive hemodynamic monitoring**: Neurosurgeries can be prolonged procedures with large volume shifts and hemodynamic changes, and neonates with brainstem dysfunction can be even more hemodynamically vulnerable. Since NIBP measurements may be technically challenging and not always accurate, arterial blood pressure monitoring can provide accurate beat to beat reading of MAP. Intra-arterial line is also useful for repeated blood sampling for blood gases, electrolytes, and haematocrit. Central venous line can be inserted for major neurosurgeries such as large NTD and can also be used for therapeutic aspiration of air from right heart in event of VAE.

(b) **Continuous monitoring of $EtCO_2$, EtN_2 and hemodynamic, precordial doppler** is advocated for early detection of VAE.

(c) **Neuromuscular monitoring** can aid in correct dosing of NMBs so as to maintain optimum intraoperative relaxation, prevent overdosing, and to help in on-table extubation.

(d) **Neuromonitoring** helps prevent inadvertent neurological damage in surgeries on spine and spinal cord and is an established monitoring technique in adult patients. Although incomplete myelination makes interpretation of intraoperative neuromonitoring (IONM) challenging in neonates, yet motor-evoked potentials, somatosensory-evoked potentials, and bulbocavernous reflex monitoring have been reported in neonates and require close communication and cooperation between the neurosurgeon and neuroanaesthesiologist, as at the time of monitoring, anaesthesiologist will need to adjust and titrate the anaesthetic technique and accordingly use short-acting agents and time the dose of muscle relaxant [71–74].

(e) **Monitoring of blood sugar, blood loss, fluid loss, and urine output** (0.5–2 mL/kg/h) should be done. The goals of fluid therapy are to maintain normovolemia, euosmolarity, euglycemia, and prevention of dyselectrolytemia. As discussed above, hypovolemia and hypervolemia are both poorly tolerated by neonates, leading to decreased cardiac output by decreasing preload and precipitating congestive heart failure, respectively. This reduces CBF and cerebral oxygenation and potentiates brain injury. Normal saline (NS) is slightly hyperosmolar and hence the preferred crystalloid during neurosurgery. However, neonates have renal immaturity and cannot handle high sodium loads and also prolonged use of NS leads to hyperchloremic metabolic acidosis. Ringer lactate (RL) is hypo-osmolar and its solitary use can precipitate cerebral edema. Moreover, neonates, especially preterm, are susceptible to hypoglycemia and require glucose supplement. All these factors have to be borne in mind to guide the fluid therapy. Glucose infusion should be given at 5–6 g/min to maintain normoglycemia and RL is used for maintenance and losses.

Craniotomies can cause significant blood loss; prior estimation of the patient's blood volume is essential in determining the amount of allowable blood loss and the time to transfuse blood. Brainstem manipulations can cause cardiorespiratory disturbances that should be immediately communicated to the neurosurgeon.

40.8.12 Lowering ICP

Raised ICP due to brain edema can be managed by:

- Increasing the depth of anaesthesia.
- Ensuring adequate muscle relaxation.
- Slight head elevation.
- Hyperventilation (moderate hypocapnia).
  Diuretics - Osmotic diuretic mannitol in a dose of 0.25–1.0 g/kg IV (it raises serum osmolality by 10–20 mOsm/kg), or Furosemide 0.1 mg/kg.

40.8.13 Prevention of VAE

Neonates are more vulnerable to the hemodynamic impact of VAE because the same amount of air enters a relatively lower circulatory volume than adults. Posterior fossa tumours are operated in prone positions and carry the risk of VAE. Although TEE and precordial doppler are most sensitive for diagnosis of VAE, $EtCO_2$ and EtN_2 are usually used for intraoperative diagnosis. Neonatal miniaturized TEE probes are available, but intraoperative use has not been documented in neurosurgery, though it was found successful in a neonate undergoing laparoscopic repair of duodenal atresia [75].

40.8.14 Extubation

Presence of normothermia and hemodynamic stability are prerequisites before considering extubation. On-table extubation followed by a focussed neurological examination has advantages of less sympathetic response related to retained ETT and lower ETT-related complications. Extubation should be smooth, avoiding coughing and bucking, without fluctuations in hemodynamic parameters, and maintaining normocapnia. Lignocaine 1.5 mg/kg can be used to block response to extubation. Residual NM blockade is reversed using glycopyrrolate and neostigmine (50μg/kg). Once the neonate is conscious and responsive, moving all limbs which are not neurologically involved and NM recovery has been ascertained, on-table extubation can be performed. Neonates who have hemodynamic instability, hypothermia, and metabolic disturbances should be electively ventilated until these disturbances are corrected. Neonates with poor preoperative level of consciousness are also difficult candidates for early extubation. A close communication with neurosurgeon to know the status of brain compliance, cerebral oedema, and brainstem handling is important before deciding for extubation. Neonates with brainstem dysfunction can undergo elective ventilation till the brainstem and lower cranial nerve functions improve.

40.8.15 Delayed Awakening

Delayed awakening in neonates could be because of metabolic factors such as hypoglycemia, hypothermia, hypocalcemia, dyselectrolytemia, or residual effect of anaesthetic drugs. Once the metabolic factors have been ruled out and neonate has still not regained consciousness even after prolonged interval from last anaesthetic drug dosage, neurological cause must be determined by using NCCT of brain.

40.8.16 Postoperative Management

Neonate should be nursed in HDU with ambient temperature maintained. O2 supplementation should be provided. Meticulous monitoring of cardiorespiratory parameters should be continued and with special vigil for occurrence of apnea in extubated neonates. Irregular respiration, apneic spells, and sleep apnea are more common in neonates with ACM and brainstem involvement. Neurogenic stridor can aggravate in postoperative period in neonates with lower cranial nerve dysfunction, necessitating re-intubation. Incomplete resolution of lower cranial nerve dysfunction, despite surgical correction, can also occur and these neonates usually require tracheostomy and have poor prognosis. Postoperative analgesia can be provided using paracetamol and short-acting opioids. Antiepileptics should be continued to prevent seizure activity. Drugs that may be used for control of seizures are phenobarbitone or phenytoin (20 mg/kg loading followed by 5 mg/kg/day) or levetiracetam (50 mg/kg loading followed by 40 mg/kg/day) [76].

40.9 Some Special Concerns in Particular Neurosurgical Procedures

40.9.1 Ventriculoperitoneal (VP) Shunt Insertion

Apart from the general concerns imposed by neonatal age, neuropathology and neurosurgery as discussed above, challenges during shunt insertion are prevention of further increase in ICP and maintenance of CPP. Due to the raised ICP, neonates who have vomited may be dehydrated and require rehydration. Raised ICP also slows gastric emptying and increased risk of aspiration, and cRSI should be considered. A large head poses problems during laryngoscopy and intubation and mandates preparedness with difficult airway cart and rescue plans. Also, surgical position requires head extension and lateral rotation that can cause accidental extubation, kinking, and signs of brainstem compression. Tunnelling of the shunt is a noxious stimulus; adequate depth of anaesthesia and adequate analgesia should be maintained at this point. Exposure of head and abdomen can lead to heat loss and hypothermia; measures for dealing with the same must be taken. Rapid drainage of

CSF from the shunt should be avoided to prevent arrythmias and hemodynamic disturbances. The complications of shunt insertion are listed in Box 40.2 [77].

Box 40.2 Complications of Shunt Insertion

- Allergic reaction to shunt material
- Hematoma formation
- Infection
- Migration into pleural cavity or heart
- Shunt fracture
- Shunt occlusion
- Valve malfunction
- Disconnection
- Over-drainage
- Outgrown shunt
- Bleeding - into subdural space/ventricles/brain parenchyma
- Injury – bowel, abdominal viscera

Neonates are particularly at high risk of complications of VP shunt, especially infection and malfunction, because of relatively thin skin, nutritional difficulties, delayed wound healing, and more proteinaceous CSF and to injury to bowel or other abdominal viscera due to the fragility of these tissues. Migration of the catheter can result in penetration into the pleural cavity or heart. Rapid decompression of large ventricles can cause bleeding into the subdural space, ventricles, or brain parenchyma. Thus, only moderate amounts of CSF are removed at the time of surgery, and patients are nursed flat immediately postoperative. Use of higher-pressure valves and programmable valves is useful [78–80].

40.9.2 Neonatal Brain Tumour Resection

Craniotomy for total or partial resection of neonatal brain tumour is a major surgery with prolonged duration in neonate. The risks include hypothermia, hypoglycemia, fluid shifts, and massive blood loss. Neonatal brain tumours are usually infratentorial that mandate use of prone position. Intracranial compliance is poor and measures to relax the brain by decreasing ICP need to be implemented. Adequate depth of anaesthesia, hyperventilation, mannitol (0.5–2 g/kg), and furosemide (0.5 mg/kg) are the strategies used for reducing ICP. TIVA is preferred over inhalational agents for maintenance of anaesthesia. Antiepileptics should be continued in the perioperative period. Posterior fossa tumours are operated in prone positions and carry the risk of VAE. Continuous monitoring of $EtCO_2$, ETN_2, haemodynamic parameters, and if available, precordial doppler aids in early diagnosis of this lethal condition. Brainstem manipulations can cause cardiorespiratory disturbances that should be immediately communicated to the neurosurgeon. Craniopharyngiomas occur in the sellar region and may be associated with endocrine disturbances, requiring steroid supplementation. They may develop diabetes insipidus (SIADH) in the postoperative period requiring replacement therapy [81].

40.9.3 Encephaloceles Repair

Encephaloceles occur due to failure of fusion of anterior neuropore during primary neurulation phase of embryogenesis, leading to protrusion of meninges and brain tissue mostly occurring in occipital and frontoethmoidal regions. Occipital encephaloceles are most common (70%) and usually contain meninges alone (meningocele) or with occipital lobes (meningoencephalocele). Ventricles may also be present along with brain tissue and meninges (meningoencephalocystocele). Occasionally, there may be other components of posterior fossa such as brainstem, cerebellum, intracranial vessels, and rarely torcula. The contents of the encephalocele along with the other associated congenital lesions determine the postoperative outcome [82].

Anaesthetic management poses several challenges to the anaesthesiologist. Apart from being associated with hydrocephalus, some of these can be of sizes even larger than the neonates' head and are called **giant occipital encephalocele**. The presence of a large occipital mass, restricted neck movements, short neck, co-existent hydrocephalus, and micrognathia imposes challenges in securing and maintaining the airway [83, 84]. Positioning the neonate for induction of anaesthesia needs care to prevent its rupture. Placing the encephalocele in a doughnut and raising the lower body to the same height can be done. Alternatively, neonate can be placed in lateral position [82, 85, 86], or the head of the neonate can be suspended beyond the head-end edge of the operating table and supported on the padded lap of the anaesthesiologist sitting at the head end, height adjusted to appropriate level, or supported by an assistant. Placing the head over a gel padded Mayfield horseshoe headrest allowing the encephalocele to hang freely beneath has also been reported. Despite the use of these manoeuvres, laryngoscopy and intubation can be difficult and difficult intubation aids including video laryngoscopes, flexible fibreoptic bronchoscope, and tracheostomy tube should always be ready. Administration of muscle relaxant must be preceded by confirmation of effective BMV. Some anaesthesiologists prefer endotracheal intubation while preserving spontaneous ventilation [86, 87]. If administered, succinylcholine is the preferred muscle relaxant due to anticipated airway difficulty. Alternatively, rocuronium can be used in institutions where sugammadex for rapid reversal is available. Movement of necks while airway management can cause brainstem compression and require careful watch on the vitals of the neonate. A co-existent ACM with symptoms of brainstem compression is associated with increased incidence of perioperative cardiorespiratory complications like arrhythmias, stridor, and postoperative apnea [88].

Surgery is conducted in the prone position that requires care of the eyes, ETT, and pressure points along with ensuring unrestricted respiratory movements. Injury of torcula, if it is present as a sac content, predisposes to cerebral deep vein thrombosis and consequent cerebral damage. There may be intracranial vessels that traverse through the encephalocele to further supply the normal brain parenchyma, and removal of such sac tissue can lead to brain infarction [82].

Cerebral malformation and lack of cerebral autonomic regulation predispose to development of hypothermia in the neonate with encephalocele who already has a large head size and large exposed surface area. Sudden loss of CSF can lead to hypotension, arrhythmias, and dyselectrolytemia. Sudden decompression can also lead to sudden traction of cerebral neurons and brainstem nuclei causing instantaneous cardiac arrest [89].

Presence of associated anomalies such as congenital heart disease can impact the perioperative anaesthetic management with its own set of problems. Anaesthetic considerations include all these concerns as discussed above along with the inherent concerns of neonatal age.

40.9.4 Meningomyeloceles Repair

Meningomyeloceles repair pose challenge in positioning the neonate for intubation. The sac can be placed in a doughnut and the remaining body can be lifted to prevent sac rupture. Presence of paraparesis below the level of the lesion warrants cautious use of succinylcholine. Bladder involvement necessitates repeated urinary catheterizations in future increasing the risk of latex allergy and hypersensitivity. Intraoperative neuromonitoring (IONM) has been deployed to minimize neurological damage during surgery and requires adjustments in doses of anaesthetic drugs and use of short-acting agents.

40.9.5 ACM Correction

ACM correction is present in almost all cases with MMC (>90%). The closure of NTD stops the outflow of CSF from the defect, leading to rise in ICP. The neonate must be followed up closely in the postoperative period for signs and symptoms of rising ICP as progressive hydrocephalus can occur which can worsen brainstem compression from ACM. This warrants an immediate VP shunt insertion. Posterior fossa decompression surgery is required if symptoms of brainstem decompression persist even after surgical treatment of hydrocephalus. Neonates with ACM leading to brainstem dysfunction have increased morbidity and mortality from cardiorespiratory events as mentioned above.

40.9.6 Craniosynostosis Surgery

Craniosynostosis is the premature intrauterine fusion of one or more cranial sutures, leading to abnormal skull growth. Multiple suture craniosynostosis may present with associated craniofacial abnormalities such as Apert's syndrome and Crouzen's syndrome. Surgical management includes strip craniectomy which should be carried out between 3 and 6 months of life to get the best result. Currently, endoscopic

suturectomy for single suture involvement is the choice to prevent morbidities of open suturectomy such as blood loss and better wound healing [51]. The management of multiple suture craniosynostosis requires multidisciplinary approach for reconstruction of face with orbital advancements. Anaesthetic concerns include a difficult airway, sudden and massive blood loss, VAE, postoperative facial edema, and possible need for postoperative ventilation.

40.10 Neuroendoscopic Procedures

Neuroendoscopic procedures include ETV, placement of VAD, septostomy, shunt placement for multiloculated hydrocephalus, transaqueductal stenting for isolated fourth ventricle, and lavage for ventriculitis or IVH.

Schulz et al. stated that despite their fragility, neuroendoscopic procedures may play an important role in the treatment of disturbed CSF dynamics in preterm and term newborn infants and may be curative in few conditions like isolated lateral ventricle, isolated fourth ventricle, CSF diversion in multiloculated hydrocephalus, ventriculitis, and IVH [90, 91].

Preterm neonates have high incidence of IVH, post-haemorrhagic ventricular dilation (PHVD), and PHH (post-haemorrhagic hydrocephalus). Management options include placement of EVD (external ventricular drain), endoscopic ventricular lavage, VAD, and ventriculo-subgaleal shunts [35–38]. Early removal of blood degradation products and residual haematoma via endoscopic ventricular irrigation is feasible and safe for the treatment of PHH with the benefit of significantly lower shunt rates and fewer complications such as infection and development of multiloculated hydrocephalus compared to conventional CSF diversion techniques [92–94].

Currently, endoscopic procedures for single suture involvement craniosynostosis are being carried out to prevent morbidities of open suturectomy. Endoscopic strip craniectomy is minimally invasive technique that offers the advantages of decreased blood loss and improved wound healing. This procedure has been carried out successfully in infants with age as low as 4–5 weeks [50, 51].

ETV may not be feasible in neonates with small-sized ventricles. Although ETV carries low morbidity than open surgery, its use in treatment of hydrocephalus in neonates remains controversial [95, 96].

Endoscopic procedures are conventionally performed in the OT under GA; their duration may vary, but the anaesthetic technique employed should be such that it enables early awakening at the end of the surgery or procedure [97].

Anesthetic implications of neuroendoscopic procedures in neonates include:

- Wide variation in duration of the procedure.
- Hypothermia and bradycardia induced by administration of cold irrigation solution.
- Sudden dilation of ventricles by the irrigating fluid can raise the ICP stimulating Cushing reflex with refractory hypertension and bradycardia.

- Fenestration through a thickened floor can also cause bradycardia.
- Rarely, haemorrhagic complications can occur that might necessitate conversion to open craniotomy.
- Arrhythmias and neurogenic pulmonary edema following acute intracranial hypertension due to manipulation of the floor of the third ventricle and lack of egress of irrigation have been reported in children during ETV. However, efficacy of ETV in neonates remains controversial, and it may not be feasible with small size third ventricles [98–100].

40.11 Conclusion

Anesthetic-induced developmental neurotoxicity is known. Very low-birth-weight babies have poor and neurological and cognitive outcomes which can be worsened by cerebral injury due to cerebral hypoperfusion, metabolic derangements, co-existing disease, and surgery; aggressive management of intraoperative hypotension, hypo or hypercarbia, oxygenation, glycemia, and temperature control is warranted in neonates undergoing neurosurgery.

Technical advances in neurosurgery and sub-specialization in neonatal neurosurgery, anaesthesiology, and critical care have improved the outcome in pediatric patients with surgical lesions of the CNS; however, in neonates it is still evolving. Lack of literature and research results leaves the clinician to use ones' clinical knowledge and acquity to manage these highly vulnerable and risky patients, with equally risky surgical and anaesthetic management, for an optimum outcome.

Evidence-based management is still evolving in neonatal head and spine trauma. Fundamental knowledge of age-related differences in cerebrovascular anatomy and physiology is essential in the application of adult based head trauma protocols in neonates.

The advancement in minimally invasive surgical and monitoring techniques, IONM, anaesthesiology, and neurocritical care, in conjunction with multidisciplinary approach comprising neonatologist, pediatric neurosurgeon, and pediatric neuroanaesthesiologist, can pave way for correction of surgical lesions of the CNS in the neonatal period in the coming future. Further advances in techniques and experience will enable more surgeries to be safely performed in neonates with improved outcomes.

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