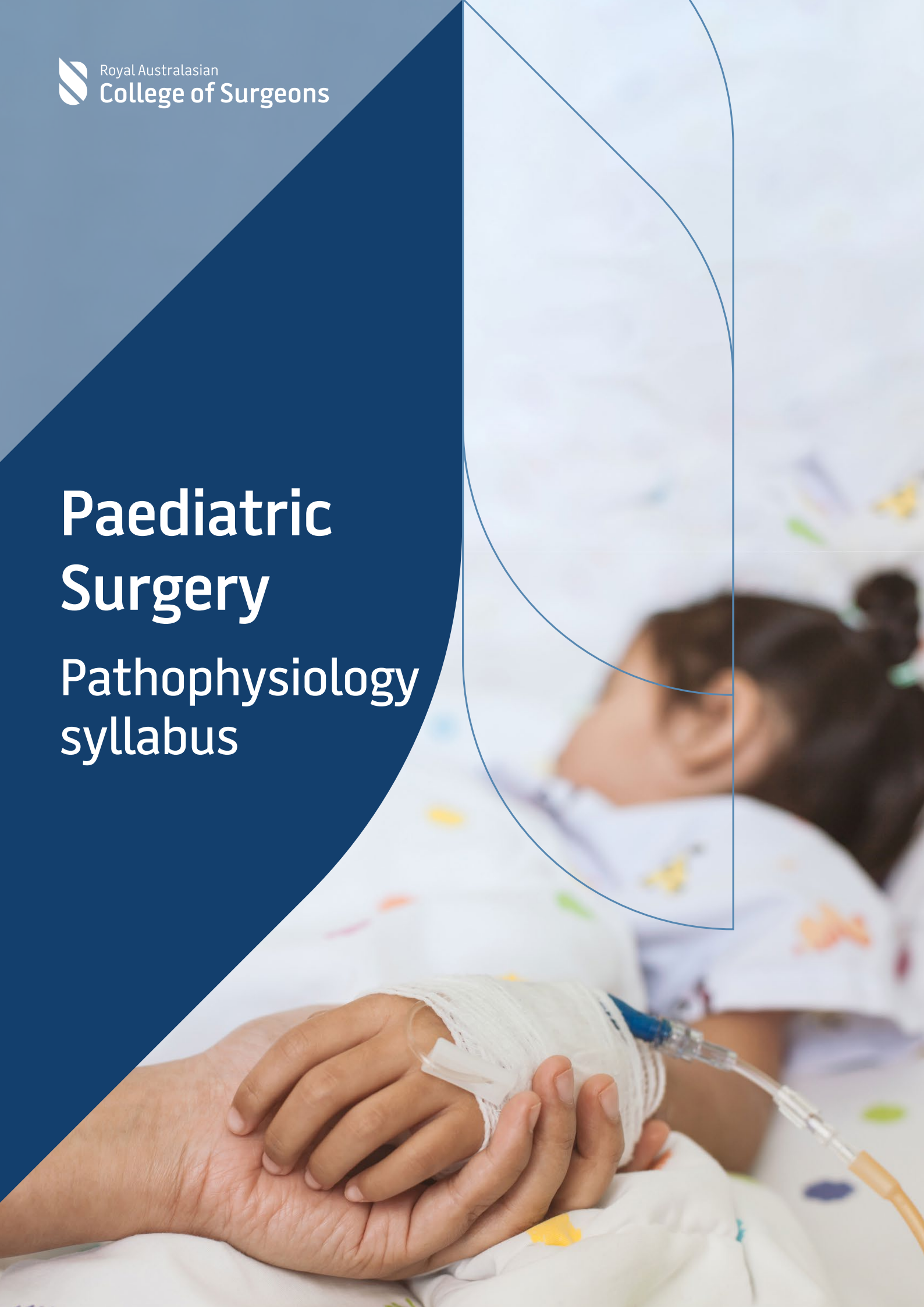


Paediatric Surgery

Pathophysiology syllabus



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Dr Parshotam Gera, WA
Dr Rebecca Cooksey, SA
Dr Sarah Giutronich, NSW

Further information

Royal Australasian College of Surgeons
250-290 Spring Street
East Melbourne VIC 3002 Australia
Telephone: +61 3 9249 1200
Web: surgeons.org

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Introduction

Pathophysiology is the study of WHY AND HOW normal physiological functioning of the body and its organs alters to a pathological state resulting in disordered functioning of the body and its organs. Several factors trigger the onset of the disease, its progression and the ultimate outcome. So, it's utterly important to have a sound knowledge of these factors while studying pathophysiology of any disease. These factors include Embryology, Aetiology, Genetics, molecular biology, pathogenesis, histopathology, risk factors, prognostic factors and associated syndromes.

Passing Pathophysiology exam is a mid-set requirement to advance to senior set. The level of knowledge achieved while preparing for the Pathophysiology exam will provide the trainee with solid foundational knowledge that they can utilise to critically analyse a clinical problem and make decisions expected of a senior set trainee.

The syllabus is divided into following sections for structure and ease of digestion. This syllabus is not exclusive or exhaustive but constitute an adequate framework to meet the learning objectives.

1. Basic science
2. Trauma and Burns
3. Neonatal surgery
4. Chest wall and Thoracic surgery
5. Gastrointestinal surgery
6. Hepatobiliary, Pancreatic and Splenic disorders
7. Oncology
8. Urology/Genitourinary surgery
9. Head and Neck surgery
10. Miscellaneous topics

On completion of this syllabus, the trainee will be able to:

Describe the embryology and normal physiological processes relevant to surgical disease.

Explain the pathophysiological mechanisms underlying Paediatric surgical conditions.

Apply the pathophysiological principles to clinical assessment and peri-operative planning.

Analyse the impact of age, associated anomalies and co morbidities on surgical risk and outcome.

Anticipate the complications based on disease mechanisms.

Integrate the pathophysiology knowledge in analysing a surgical problem and clinical decision making.

Basic science

Nutrition

- Metabolic response to stress/injury
- Macronutrients
- Micronutrients
- Enteral and parenteral nutrition

Anaesthetic considerations

- Anaesthetic related neurotoxicity
- Preoperative anaesthetic evaluation
- Malignant hyperthermia
- Ambulatory surgery
- Anaesthesia in high-risk population
 - Upper respiratory tract infection
 - Trisomy 21
 - Preterm infant
 - Anterior mediastinal mass
 - Congenital heart disease
- Effects of pneumoperitoneum and Pneumothorax on different organ systems during minimally invasive surgery

Renal physiology

- Fluids and electrolytes homeostasis
- Acid base balance
- Renal function evaluation
- Acute kidney injury
- Dialysis
- Renovascular hypertension

Coagulopathies

- Normal Hemostatic response
- Disseminated intravascular coagulation
- Thromboelastography [TEG] and Rotational thromboelastometry [ROTEM]
- Haemophilia, Von Willebrand disease and Thrombocytopenia
- Surgical implications of Sickle cell disease
- Blood and blood products

Extracorporeal membrane oxygenation

- Principles
- Types
- Indications and contra indications
- Complications and long term sequelae

Foetus as a patient

- Routine antenatal screening tests and specific diagnostic tests when an anomaly identified
- Conditions potentially benefit from foetal surgery
- Inclusion and exclusion criteria for foetal surgery
- Hydrops foetalis
- Principles of EXIT strategy

Trauma and burns

Burns

- Epidemiology of paediatric burns, patterns of injury and prevention strategies
- Classification of burns based on the depth of burn injury
 - contributing factors
 - assessment and sequelae of differing depths
- The local response to burn injury
 - Jackson's zones of burn wound and its clinical implications.
 - Mechanism of circulatory impairment in a circumferential burn
 - Mechanism of respiratory impairment after trunk burns
- Systemic response to burn injury
 - Circulatory
 - Cardio-pulmonary
 - Renal
 - Gastrointestinal
 - Immune
 - Metabolic
- The process of hypertrophic scarring and scar contracture including scar maturation
- Keloid and Hypertrophic scars

Inhalation injury

- Classification and Pathophysiology of Airway inhalation injury (thermal, chemical and systemic toxicity)

Chemical burns

- Classification of chemical burns
- Mechanism of injury in different types of chemical burns

Electrical burns

- Types of electrical burns
- Pathophysiology of electrical burns
- Compartment syndrome

Shock

- Classification of shock (distributive, hypovolaemic, cardiogenic and obstructive)
- Pathophysiology of different types of shock
- Physiologic response to shock immediately and over time, both treated and untreated

Abdominal trauma

- Mechanism of injury and patterns of abdominal injury in childhood.
- Anatomical and physiological characteristics/differences in children relevant to trauma
- Short term and long-term sequelae of traumatic injury to abdominal organs.

Neck trauma

- Patterns of injury in paediatric neck trauma
- Three zones and associated anatomic considerations in relation to neck trauma

Chest trauma

- Patterns of thoracic injury in childhood and how these patterns of injury relate to differences in thoracic anatomy in children.
- Classification and pathophysiology of chest trauma – Chest wall/pleura/lungs/Mediastinum, including:
 - Pneumothorax
 - Haemothorax
 - Pulmonary contusion

- Rib fractures/flail chest
- Cardiac injury
- Tracheo-bronchial injury
- Mediastinal injury

Soft tissue/limb trauma

- Recognise types of soft tissue injury that are associated with different mechanisms of trauma and associated injuries (e.g. seatbelt sign)
- Local and systemic effects of crush injury to a limb

Neonatal surgery

Neonatal Physiological Principles Relevant to Surgery

Trainees should demonstrate an understanding of:

- Cardiovascular transition from foetal to neonatal circulation
 - Pulmonary adaptation at birth and surfactant physiology
 - Thermoregulation and metabolic demands
 - Fluid compartments, electrolyte balance, and acid–base regulation
 - Renal immaturity and implications for fluid and drug handling
 - Hepatic immaturity, bilirubin metabolism, and coagulation
 - Immune system immaturity and infection risk
 - Haematological physiology, including anaemia and haemostasis
 - Anaesthetic and perioperative implications in the management of a neonate
- Hypoplastic left colon
 - Necrotising enterocolitis
 - Meconium-related disorders
 - Abdominal wall defects and associated physiological derangements
 - Anorectal malformation
 - Classification
 - Associated syndromes
 - Prognostic indicators
 - Congenital diaphragmatic hernia
 - embryology of diaphragm
 - Classification
 - antenatal risk factors and implications
 - pathophysiology of pulmonary hypoplasia and pulmonary hypertension
 - Oesophageal atresia and tracheo-oesophageal fistula
 - Embryology
 - Classification
 - Associations including syndromes and genetic conditions
 - Pathophysiological implications of ventilation

Surgical conditions affecting Neonates

- Embryological development of the gastrointestinal tract- oesophagus to anus
- Duodenal and intestinal atresia
- Gut rotation
 - Embryology
 - Anomalies of gut rotation
- Hirschsprung disease
 - Embryology
 - Genetics
 - Histology
 - Pathophysiology
 - Hirschsprung enterocolitis

Chest wall and thoracic surgery

Lung and Oesophagus

- Embryology of foregut development
- Stages of lung development with timeline and bronchopulmonary structures formed in each stage
- Embryological theories of development of the spectrum of congenital cystic lung lesions, this framework should cover both intra and extra pleural lesions.
- Classification of congenital Bronchopulmonary malformations
- Pathophysiology and long-term sequelae of Bronchopulmonary malformations
- Foregut cysts - Embryology and pathophysiology
- Caustic injuries of Oesophagus
- Barrett`s oesophagus
- Achalasia Cardia – Aetiology and pathophysiology
- Foreign body Oesophagus – sites of physiological narrowing and likely impaction, pathological consequences of impaction
- Oesophageal strictures

Chest wall anomalies

- Aetiology and pathophysiology of all chest wall anomalies including but not limited to Pectus excavatum, Pectus carinatum and Poland syndrome
- Long term functional implications of these abnormalities

Empyema Thoracis

- Causative organisms
- Patho- physiological sequence of events in the formation of empyema, and the long-term complications associated with this disease process.
- Stages of empyema thoracis
- Pathophysiology of Necrotising pneumonia
- Lung abscess

Bronchiectasis

- Classification
- Pathophysiology of bronchiectasis
- Causes, possible associations and the microbiology of the condition.

Intrathoracic masses

- Causes of intra thoracic masses in children.
- •Patho-physiological consequences of intra-thoracic masses and their implications, including peri-operative concerns.

Spontaneous pneumothorax

- Pathophysiology of spontaneous pneumothorax
- Risk factors for and conditions associated with spontaneous pneumothorax

Pneumomediastinum

- Aetiology
- Pathophysiology

Chylothorax

- Aetiology of Chylothorax
- Composition of Chyle
- Pathophysiology of Chylothorax and its systemic consequences.

Gastrointestinal tract

- Detailed understanding of development of GI tract – Both normal and aberrant
- Gastroesophageal reflux
 - Anatomical and physiological barriers against reflux
 - Natural history of reflux in infants
 - Risk factors for reflux
- Hypertrophic pyloric stenosis
 - Proposed theories
 - Associated dyselectrolytaemia
 - Classic metabolic derangement
 - Sequela of inadequate correction
- Gastric volvulus
 - Types with clear understanding of each type
 - Triad of symptoms
- Intussusception
 - Types
 - Causes
 - Pathophysiology and complications
- Alimentary tract duplications
 - Embryology
 - Classification based on location
 - Pathological consequences
- Meckel`s diverticulum
 - Morphological types of omphalomesenteric duct remnants
 - Complications
 - Meso-diverticular band
- Appendicitis – Etiology, pathophysiology and complications
- Inflammatory bowel disease
 - Etiology
 - Pathophysiology
 - Macro and microscopic appearances
 - Extra intestinal manifestations
- Acquired anorectal conditions
 - Perianal abscess
 - Fistula in Ano
 - Fissures
 - Rectal prolapse
- Short gut – Causes and pathological consequences, Normal bowel length at different ages
- Constipation and faecal incontinence – medical and surgical causes; Physiology of gut motility and faecal continence mechanism.
- GI bleeding – Potential sites of bleeding and relate this to common pathologies seen in children of different age groups
- GI polyps – classification, pathological consequences and underlying genetic disorders
- Chylous ascites
 - Aetiology
 - Composition of Chyle
 - Pathophysiology of Chylous ascites and its systemic consequences.

Hepatobiliary, pancreatic and splenic disorders

Hepatobiliary

- Embryology
 - Normal embryological development of the liver, gallbladder and bile ducts
 - Developmental anomalies of the liver, gallbladder and bile ducts and the potential clinical consequences
- Physiology
 - Normal physiological functions of the liver in infants and children
- Neonatal jaundice
 - Physiological jaundice in neonates, and how this differs from pathological jaundice
 - Classification
 - Classify causes of neonatal jaundice and demonstrate understanding of the implications for investigation and management
 - Brief knowledge of medical causes of conjugated hyperbilirubinaemia
- Biliary atresia
 - Classification – Anatomical and clinical classification
 - Aetiology
 - Etiological theories for development of biliary atresia and the potential impact on presentation and outcomes
 - Knowledge of potential associations (conditions and syndromes)
 - Pathophysiology
 - Natural history of biliary atresia if untreated
 - Typical pathological features of biliary atresia on liver biopsy
 - Prognostic indicators
- Cholelithiasis
 - Aetiology of gallstones in children
 - Classification
 - Classification of gallstones and will be able to outline specific associations or risk factors for each type
 - Pathophysiology of biliary colic, cholecystitis, cholangitis and gallstone pancreatitis

- Choledochal cyst
 - Epidemiological factors and incidence of choledochal cyst in different populations
 - Aetiology – Theories for choledochal cyst development and the proposed mechanisms
 - Classification (Todani)
 - Pathophysiology
 - Understanding of the pathophysiological processes leading to the clinical manifestations of choledochal cysts
 - Long-term issues related to choledochal cysts
- Inspissated bile syndrome – Aetiology and pathophysiology
- Spontaneous biliary perforation – Aetiology and pathophysiology
- Portal hypertension
 - Classification of portal hypertension in children (pre-hepatic, hepatic, post-hepatic), Common causes of portal hypertension in children
 - Pathophysiology
 - Pathological consequences of portal hypertension in children, specifically the development of varices and how this may manifest
 - Sites of porta systemic shunts
- Ascites
 - classification of ascites in children
 - Pathologies leading to ascites in children and how these might be differentiated

Pancreatic disorders

- Embryology
 - Embryological development of the foregut and the pancreas
 - Anatomical/embryological variants in development of the pancreas and the potential clinical consequences
- Physiology
 - Normal functions of the pancreas
- Pancreatitis
 - Classify pancreatitis in children and outline the common causes of

- pancreatitis and how this differs from adult patients
- Pathophysiological processes that lead to development of pancreatitis, and how this results in the clinical manifestations of pancreatitis
- Hereditary pancreatitis and the specific gene mutations that are described
- Hyperinsulinism
 - Classification of congenital hyperinsulinism and implications for management
 - Gene mutations leading to congenital hyperinsulinism (focal and diffuse)
 - Clinical manifestations of congenital hyperinsulinism

Splenic disorders

- Embryology
 - Embryological development of the spleen
 - Anatomical/embryological variants in development of the spleen and the potential clinical consequences
- Physiology
 - Normal function of the spleen during foetal development and childhood
- Demonstrate understanding of the different conditions that may require splenectomy during childhood
- Consequences of asplenia in childhood
 - Causes and pathophysiology of overwhelming post-splenectomy infection (OPSI) including specific organisms/importance of immunisation
 - Potential findings on blood film after splenectomy

Oncology

General oncologic considerations

- Familiarity with chemotherapeutic agents, their mechanisms of actions and their side effects that will have implications for surgical patients. (e.g. tumour lysis, typhilitis, Bone marrow suppression etc.)
- Familiarity with radiotherapy techniques, their side effects and surgical issues relating to the use of radiotherapy.
- Familiarity with nuclear medicine techniques of diagnosis and treatment in relationship to paediatric oncology.
- General principles of flow cytometry, cytogenetics and immunohistochemistry
- Hereditary cancer syndromes of relevance to paediatric practice and inherited gene mutations.
- Tumour markers relevant to Paediatric malignancy.
- Genetic mutations and translocations that are of diagnostic, prognostic or therapeutic importance i.e. EWS/FLY Ewings or FOX/PAX translocations for Rhabdomyosarcoma.
- Graft versus host disease.

Vascular Anomalies

- Commonly accepted classification of vascular malformations, and advantages of this system over previous nomenclatures.
- Current theories of pathogenesis of all types of vascular anomalies, their development and natural history.
- Embryology of lymphatic system, classification of lymphatic malformation and spectrum of clinical presentations depending on the site of occurrence.
- Lymphedema
- Vascular syndromes seen in the neonatal period
- Types of Haemangiomas, Pathophysiology and natural history, including that of Hepatic Haemangioma
- Venous vascular malformations – Pathophysiology and natural history
- AV Malformations – Clinical staging and natural history
- Combined vascular malformations – Different syndromes and clinical

manifestations

Rhabdomyosarcoma

- Associated syndromes
- Histological subtypes
- Tumour biology and prognostic implications
- COG risk group stratification
- Different tumour types in specific locations, different age groups and effects on clinical prognosis and tumour behaviour.

Non-Rhabdomyosarcoma soft tissue tumours – Including PNET and fibromatosis group

Neuroblastoma

- Origin of neuroblastoma and the potential sites of occurrence.
- International Neuroblastoma pathology classification, molecular biology and risk stratification
- Metabolic tumour by-products and relate this to clinical presentations and their consequences.

Nephroblastoma (Wilms tumour) & Congenital Mesoblastic nephroma

- Pathogenesis and histological subtypes
- Various syndromes and associated genetic abnormalities
- Nephrogenic rests and Nephroblastomatosis
- Types Mesoblastic nephroma, its similarities and differences to Wilms tumour
- Other childhood renal tumours and their key differences to Wilms tumour

Gonadal tumours

- Classification of Gonadal tumours with respect to the cell lines of origin
- Detailed knowledge of Germ cell tumours of testis and ovary
- Knowledge of other benign and malignant gonadal tumours, their presentation and diagnosis in relationship to their histologic type and cell function.

Lymphoma

- Histological classification of

lymphomas.

- Clinical situations in which lymphoma can present as or complicate general surgical/urological diseases.
- Staging of lymphoma.

Teratoma/ Sacrococcygeal teratoma

- Embryologic origin of teratoma, sites of occurrence and clinical manifestations
- Sacrococcygeal Teratoma – classification and clinical implications of giant Sacrococcygeal teratoma
- Histological features of mature, immature and malignant teratoma.
- Tumour markers and their role in diagnosis, prognosis, and monitoring for teratomas.

Hepatic tumours

- Primary malignant tumours of liver
- Hepatoblastoma – associated conditions, histological subtypes, tumour biology and cytogenetics
- Hepatocellular carcinoma and Embryonal sarcoma
- Benign Hepatic tumours

Pancreatic tumours

- Classify the tumours of the pancreas
- Clinical manifestations

Intrathoracic masses

- List the causes of intra thoracic masses in children
- Patho-physiological consequences of intra-thoracic masses and their implications, including peri-operative concerns.

Thyroid & Parathyroid

- Nodules; Malignancy; Thyroiditis; Goitre; Ectopia
- Etiology and familial factors in paediatric thyroid disease
- MEN – genetic causes, subtypes and relevant associated conditions in the subtypes

Adrenal tumours

- Classification
- Clinical manifestations
- Associated syndromes

Urology and genitourinary system

-
- Embryology of upper urinary tract, lower urinary tract and external genitalia.
 - General considerations
 - Tubular function and transport disorders
 - Acid base regulation
 - Dyselectrolytaemia in renal disease
 - Post obstructive diuresis
 - Foetal urine and amniotic fluid, Urinary biomarkers of renal damage
 - CAKUT – Detailed knowledge of these conditions including relevant embryology is expected
 - Renal Parenchymal anomalies – Renal agenesis, Renal hypoplasia, Renal dysplasia, MCDK, Cystic renal disease
 - Anomalies kidney position and shape – Horseshoe kidney, Ectopic kidney, Crossed fused ectopia
 - Renal collecting system and ureteric anomalies – Hydronephrosis, PUJ obstruction, VUJ obstruction, Duplex system, Megaureter, Ureterocele
 - Bladder and Urethral anomalies - PUV, Bladder exstrophy epispadias complex, Anterior urethral obstruction, Urethral duplication
 - Urinary tract infections
 - Pathogenesis
 - Understanding the difference between upper and lower urinary tract infection
 - Host and pathogen interactions in UTI.
 - Awareness of how abnormalities and dysfunction of renal tract contribute to UTI.
 - Vesicoureteric reflux
 - Classification
 - Pathogenesis and grading
 - Natural history
 - Reflux nephropathy.
 - Chronic kidney disease
 - Aetiology
 - Staging and complications
 - Indication for renal replacement therapy and transplantation.
 - Megacystis – causes and clinical implications
 - Bladder diverticulum
 - Types
 - Causes
 - Pathophysiology
 - Urethral Stricture Causes and Classification
 - Hypospadias - Embryology of penis and urethra in relation to development of hypospadias
 - Prune Belly Syndrome
 - Embryological Theories
 - Triad
 - Pathophysiology
 - Urinary Tract Calculi - Types of Stones and Risk Factors for Stone Formation
 - Discuss Pathophysiology of Haematuria Renal, Urethral, Ureteric, Bladder, and Urethral Origin
 - Bladder and Sphincter Innervation, Physiology of Micturition
 - Anatomical, Embryological, and Behavioural Factors Contributing to Lower Urinary Tract Function and Dysfunction
 - Neurogenic bladder – Aetiology and classification
 - Non neuropathic neuropathic Bladder [formerly known as Hinman Syndrome]
 - Nocturnal Enuresis
 - Types
 - Pathophysiology
 - Classification of disorders of sexual differentiation with underlying pathophysiology.
 - Precocious puberty
 - Balanitis Xerotica Obliterans [lichen sclerosis].
 - Testis
 - Embryology
 - Normal descent of testis
 - Histology of undescended testis.
 - Acute scrotum, differential diagnosis. Testicular torsion.
 - Varicocele - Aetiology, classification and pathology
 - Ovary - Pathological conditions that occur antenatally during childhood and adolescence, including neoplasms, cystic conditions, and torsion.
 - Labial adhesions. Aetiology and clinical implications.
 - Introital masses - Differential diagnosis.
 - Uterine and vaginal anomalies.

Head and neck

- Branchial apparatus
 - Embryology and its remnants.
 - Course and location of internal opening of different types of Branchial cysts
- Thyroglossal cyst
 - Embryology and location
 - Pathology of thyroglossal cyst.
- Preauricular sinus and cysts.
- Dermoid and epidermoid cysts.
- Torticollis
 - Causes
 - Pathological basis
 - Natural history of sternomastoid tumour
- Cervical lymphadenitis – Causes and pathophysiology including atypical mycobacterial infections.
- Tongue tie.

Miscellaneous conditions

- Congenital melanocytic nevi
 - Classification
 - Histology and malignancies.
- Acquired melanocytic nevi – risk factors
- Pathological features and types of abnormal nevi.
- Spitz nevi.
- Melanoma – risk factors and prevention.
- Umbilicus
 - embryology and natural history of umbilical hernia
 - Causes of umbilical discharge, their embryological basis and clinical consequences.
- Epigastric hernia, spigelian hernia, lumbar hernia, and femoral hernia.
- Inguinal hernia and hydrocele – risk factors, pathogenesis of hernia/hydrocele, types of hydroceles.
- Breast
 - Embryology
 - Hormonal effects on it
 - List causes of breast enlargement in children
 - Gynaecomastia – list causes
- Obesity
 - Definition
 - Complications of obesity
- Palmar hyperhidrosis - causes and use of this knowledge to understand its treatment.
- Hidradenitis suppurativa - pathology and its natural history.
- Disorders of toenail - Ingrowing toenail.
- Benign soft tissue tumours
 - pilomatrixoma, juvenile xanthogranuloma, nodular fasciitis, and hamartomas.
- Pilonidal sinus
 - Pathogenesis
 - Progression and long-term outcomes.
- Embryology and physiology of adrenal gland.
- Thyroid and parathyroid pathologies in childhood and adolescence.
- List causes of adrenal masses and clinical manifestations.
- Precocious puberty.
- Infections
 - Wound classification
 - Cellulitis
 - Necrotizing fasciitis
 - Tetanus
 - Hydatid cyst
 - Molluscum contagiosum
 - Atypical Mycobacterial infections
 - Cat scratch disease.

