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**Pathophysiology Examination Syllabus**

SET in Paediatric Surgery

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**SET PROGRAM IN PAEDIATRIC SURGERY – PATHOPHYSIOLOGY EXAMINATION**

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### **INTRODUCTION**

The syllabus is divided into sections for ease of digestion and structure.

There is overlap as certain conditions will fall under different headings for example gastroschisis has issues of relevance to embryology and nutrition and growth.

The Trainee is invited to utilise this syllabus as a guide to the important areas of study, but may wish to construct their study program on another basis.

This syllabus assumes a background level of knowledge consistent with a passing grade in the Generic Surgical Sciences Examination (GSSE), and the Specialty Specific Paediatric Anatomy and Embryology Examination. The learning objectives described here will build on that base.

The Specialty Specific Pathophysiology Examination is a Mid-SET requirement for advancement to Senior-SET.

Whilst independent clinical decision making is the standard to develop in Senior SET (especially for the FEX), the level of knowledge expected to be achieved for this exam will provide the Trainee with the basic science, epidemiology, pathology, pathophysiology etc. to inform the discussions and decisions expected of a Senior SET/ Graduate of the program.

Therefore, while these learning objectives are not specified in terms of clinical management; clinical scenarios may be employed as a vehicle for discussion or questioning and as a means of demonstrating relevance of the material for the practising paediatric surgeon.

### **OUTLINE**

- 1: Embryology/Developmental
- 2: Neonatal
- 3: Fluids/Nutrition/Growth
- 4: Trauma/Burns
- 5: Infection/inflammation
- 6: Neoplasia
- 7: Other Acquired abdominal disorders
- 8: Genito-Urinary
- 9: Skin/Subcut/bodywall/extremities

In addition to the topics and learning goals there is advice on recommended reading. This is not exclusive or exhaustive but should constitute an adequate grounding to meet the learning objectives.

**1: Embryology/Developmental**

The Trainee will be able to apply their knowledge of the embryology of relevant organ systems to clinical issues that arise in the perinatal period and later in life.

The Trainee will be able to discuss developmental issues that present as clinical issues in paediatric surgical practice.

The following list of conditions should be understood in detail as they constitute major areas of paediatric surgical practice. Other diagnoses and derivation from normal growth and development may also be relevant.

**Head and Neck:** The Trainee will demonstrate understanding of the embryologic process of head and neck development as it pertains to the listed conditions in detail. The Trainee will recall important facets of other embryologic process and abnormalities in the head and neck region that they may encounter in paediatric surgical practice.

**Branchial apparatus remnants**

Elucidate embryological basis for branchial apparatus remnants. Define the pathological findings and anatomical variants.

**Thyroglossal remnants**

Elucidate embryological basis for thyroglossal remnants. Define the pathological findings and anatomical variants.

**Dermoid cyst head and neck**

Describe embryological basis, pathological findings and natural history.

**Pre-auricular sinuses and cysts**

Describe pathogenesis and embryological basis

Conditions to have knowledge in less detail; Cleft lip/palate

**Body wall:** The Trainee will demonstrate and understanding of the embryological development of the body wall. The Trainee will discuss the implications of deviations from normal development and how these result in the separate recognized conditions of body wall abnormalities. The Trainee will be able to explain the associations seen with different abnormalities of body wall development (See also Fluids/Nutrition/Growth).

**Abdominal Wall**

Explain the embryology and anatomy of the abdominal cavity.

Explain the embryology and anatomy of the relationship of the gut to the abdominal wall.

Describe Explain when and how gastroschisis occurs. List the associated anomalies that may occur.

Describe Explain when and how exomphalos occurs. List conditions that maybe associated with exomphalos

Discuss the body wall abnormality seen in Triad (Prune Belly) Syndrome (see also Urinary tract)

**Chest wall**

Explain the different types of chest wall deformity, including Poland's syndrome

Describe the natural history of chest wall deformities and their physiological implications

**Umbilicus**

Explain the embryology and anatomy of the umbilicus. Discuss clinically important implications of this.

Describe the natural history of umbilical hernia untreated

Explain the causes of umbilical discharge and the embryological basis of each

Describe the clinical consequences of omphalitis with reference to the embryology of the umbilicus

**Epigastric hernia**

Describe the anatomy of epigastric hernia and explain how epigastric hernia produces symptoms

**Inguino-scrotal**

Describe the anatomy of the inguinal region, including the relationship of the structures within the spermatic cord.

Describe the embryology of the inguinoscrotal region and why hernias and hydroceles may occur

Classify groin hernias. Describe the embryology of the processes vaginalis and testicular descent.

Explain the embryology of gonadal development including descent of testis and associated anomalies of embryological development.

Explain the significance of an absent vas or a gonad in a hernial sac.

Have knowledge of other hernias of the lower abdominal wall and groin eg femoral, spigelian, lumbar etc.

**Spine**

The Trainee will recall the process of neural tube development, and list the types of neural tube defects.

**Diaphragm (CDH):** As an index condition the Trainee must have detailed knowledge of both the embryological abnormalities and the disordered physiology that is associated.

Describe the embryological theories for the development of the condition.

Describe the pathogenesis especially with regard to the development of lung hypoplasia and pulmonary hypertension.

Describe the different types of congenital hernias seen in the diaphragm.

(See also neonatal)

**Lung/Thorax:** The Trainee will have an understanding of the pathology observed in congenital lung lesions, and an understanding of the sequelae associated with them.

Discuss embryological theories for development of the spectrum of congenital cystic lung lesions, this framework should cover both intra and extra pleural lesions.

Describe the pathogenesis of the different types of congenital cystic lesions including long term sequelae.

Explain how the embryological theories of lung development relating to the spectrum of abnormalities seen with oesophageal duplications and bronchogenic cysts.

Describe the anatomy of the lungs and major airways

**GI Tract:** The Trainee will have a detailed understanding of the development (both normal and aberrant) of the whole GI tract. The Trainee will be able to discuss the range of clinical conditions associated with deviation from normal development. The Trainee will describe the physiological implications both short and long term of abnormalities of GI development (see also Fluids/Nutrition/Growth)

**Oesophagus**

Understand the embryology of foregut formation and its relationship to development of oesophageal atresia, and apply this to the various conditions of abnormal development

Discuss the types of oesophageal atresia with or without tracheo-oesophageal fistula (including H fistula) and understand how this affects presentation and prognosis

Discuss long term functional implications of these abnormalities (incl treatment related)



### **Bowel**

Describe the types of atresia that may occur in the bowel from duodenum down to rectum. Know their frequency and associations. Have an understanding of the theoretical embryological basis of each type

Describe the disorders of intestinal duplication at each site in the GI tract.

Discuss the embryological theories of intestinal duplication and associated anomalies. (incl neuro-enteric cysts)

### **Meckel's diverticulum**

The Trainee will be able to relate the specific pathological presentation to the embryologic development of the midgut.

### **Malrotation**

Discuss the process of normal intestinal rotation (incl as above association with abdominal cavity development)

Understand the types of malrotation including non rotation

### **Ano-rectum**

Discuss the embryology of ano- rectal development and the theories for the different anomalies encountered.

Describe a classification system of anorectal abnormalities for both males and females

Understand the condition of cloaca.

Know the effects on development of the surrounding tissues and apply this to describing the implications this has on future function.

**Renal tract:** The Trainee will be able to demonstrate understanding of the embryological development of the renal tract as a whole and specific deviations from normal as they affect each site of the renal tract. The Trainee will also understand the normal physiological functions of the renal tract (both antenatal and in transition to post-natal life) and the clinical implications of disordered function; this will include antenatal and postnatal effects, and renal and extra-renal effects. The Trainee will recognize that some deviations from normal function and appearance may not constitute a clinically relevant pathology.

### **Renal tract dilatation**

Explain the development of renal tract dilatation and the differential diagnosis including current theories of embryological basis. Discuss associated conditions particularly of renal tract anomalies and impact on outcome and management.

### **Multicystic dysplastic kidney**

Explain the development of the multicystic kidney and the differential diagnosis of congenital renal cystic lesions.

### **Posterior urethral valves**

Describe the embryological basis of posterior urethral valves and the associated anatomical and functional findings. Be able to discuss patho-physiology of clinical presentation both ante-natally, perinatally and delayed presentation

Analyse the effects of urinary tract obstruction on the fetus with particular reference to Potters Syndrome.

Understand the long term physiological effects of antenatal obstruction and describe features of the condition known as "Valve Bladder"

### **Triad/Prune Belly Syndrome**

Discuss the embryological theories of the development of Triad Syndrome.

Discuss the disordered function associated with this.

### **Ureterocele**

Describe the embryology and anatomy of ureterocele and the associated structural findings.

Discuss differences between single system and duplex ureteroceles

**Hypospadias**

Describe the embryology of the penis and urethra in relation to the development of hypospadias

**Preputial conditions**

Describe the embryology and natural history of prepuce

**Urinary incontinence**

Describe the embryology and anatomy of the urogenital tract and the normal development of urinary continence

**Neuropathic / neurogenic bladder**

Describe the anatomy and innervation of the bladder and sphincter and list the causes of neurogenic bladder

Classify neurogenic bladder with reference to pressure, outcome, continence, infection and potential for renal damage (uropathy)

**Vesicoureteric reflux and UTI**

Describe the embryology and anatomy of the urinary tract related to the condition of vesicoureteric reflux and the associated findings. Discuss theories of natural progression.

**Genito-urinary:** The Trainee will discuss the normal embryologic development of the uro-genital system; including sexual differentiation and formation of both male and female genital tracts. The Trainee will understand the normal physiological function of both male and female genito-urinary systems to the extent relevant to normal paediatric surgical practice.

**Labial adhesions**

Describe the aetiology of labial adhesions

Discuss any associations and clinical implications

**Disorders of sexual development**

Describe the embryology of the urogenital tracts with reference to disorders of normal development and endocrinological and chromosomal control of sexual differentiation

**Bladder Exstrophy and Epispadias**

Describe the embryology of the external genitalia, urethra and bladder neck and relate to the anatomical abnormalities of bladder exstrophy and epispadias including associated orthopaedic anomalies

Understand the additional abnormality of cloacal extrophy

**Hydrometrocolpos**

Describe embryology and anatomy of female genital tract relating to uterine and vaginal anomalies causing hydrometrocolpos List associated urinary and G.I.T. abnormalities.

**Ovarian pathology**

Discuss the ovarian pathologies that occur antenatally, during childhood and adolescence including neoplasms, chromosomal anomalies and cystic conditions. Discuss embryological development of ovaries and internal genital tract with regard anomalies including antenatal torsion

**Acute scrotum**

Classify testicular torsion and relate to the predisposing anatomical configurations.

Explain embryology of testis and renal tract and its relationship to torsion and other causes of acute scrotum.

Discuss path- physiological and anatomical basis for non-surgical causes of acute scrotum

**Varicocele**

Describe the embryological theory of development of the testis, inferior vena cava and gonadal veins as it relates to development of varicocele. Discuss post-natal pathological causes of varicocele.

**Vascular anomalies:** The Trainee will have an understanding of the range of vascular anomalies and their effects on normal anatomy and function throughout the body. The Trainee will understand the natural history of vascular lesions and the implications of this for treatment. The Trainee will describe how vascular lesions may have a functional impact on adjacent structures and tissues –both with respect to treated and untreated.

Explain the commonly accepted classification of vascular malformations.

Relate the current pathogenesis theories of vascular anomalies to their development and natural history.

Describe the embryology of the lymphatic system related to sites of appearance of lymphatic malformations

Describe the vascular syndromes seen in the neonatal period.

**Lymphoedema**

Describe the embryology and pathophysiology of the lymphatic system

Describe the embryology of the great vessels –above and below the diaphragm. Use knowledge of variation from normal development to explain clinical manifestations.

## **2: Neonatal**

The Trainee will have an understanding of antenatal findings that accompany conditions that require post-natal surgical management. The Trainee will understand the ante-natal findings that are associated with abnormal physiology and the effects on the newborn in those settings.

The Trainee will have some knowledge of the issues around ante-natal interventions for surgical conditions.

The Trainee will have an understanding of the normal physiological processes involved in the transition from ante-natal to post-natal functioning. The Trainee will demonstrate understanding of the unique management issues in the neo-natal surgical patient including fluid management, nutrition, homeostasis –including heat transfer, and issues around transfer.

The Trainee will have an in depth knowledge of the pathophysiological processes that accompany surgical disease and contribute to morbidity and mortality in the neonatal period.

The Trainee will demonstrate familiarity with ante-natal and post-natal screening. Both in terms of the normal process and the disorders that maybe detected that will have relevance to surgical patients.

**NEC:** The Trainee will understand the pathologic basis of the disease and factors that can lead to its development.

The Trainee will analyse the theories behind the development of NEC. The Trainee will apply their knowledge of the patho-physiological process in discussing the assessment and management of NEC.

### **Cystic fibrosis and meconium ileus**

Discuss pathogenesis of complicated and uncomplicated meconium ileus. Discuss other forms of meconium disease seen in the neonate. Understand the role of the stoma in meconium ileus and historically the types that have been described to manage the condition. Discuss the theoretical basis of other treatment strategies in Meconium Ileus. (see also Fluids/Growth/Nutrition, also related to embryology)

**Persistent hyper- insulinaemic hypoglycaemia of infancy:** The Trainee will describe the possible causes of persistent hyperinsulinaemia in the neonate. The Trainee will understand the pathogenesis of elevated insulin.

**Hirschsprung's disease:** The Trainee will understand the embryological theories for the development of Hirschsprung's disease and known genetic associations.

The Trainee will be able to differentiate between HD and other causes of bowel dysfunction in the neonatal period.

The Trainee will describe the pathological effects on the bowel of neuronal deficiency. The Trainee will describe the effects on gut innervation and how this can be used to make the diagnosis. The Trainee will identify the pathological findings in the bowel that are associated with Hirschsprung's disease.

The Trainee will understand the pathological basis for HD related enterocolitis.

The Trainee will discuss theories behind the contributory factors to HD related enterocolitis.

**Biliary Atresia:** The Trainee will understand normal and pathological jaundice in the neonatal period. The Trainee will describe differentiation and management of non-obstructive jaundice. The Trainee will discuss the factors that may have a role in the etiology of the disease and know any associated conditions and syndromes. The Trainee will describe the pathologic changes which occur in the biliary system with biliary atresia. The Trainee will apply their knowledge in discussion of the progression of the disease both with and without intervention. The Trainee will describe the pathological features of progressive disease. The Trainee will describe the other pathologies that are associated with jaundice in the newborn period and early childhood.

**Torticollis:** The Trainee will describe the pathological basis and causes, and the natural history of Sternomastoid tumour.

**Infantile Hypertrophic Pyloric Stenosis:** The Trainee will describe the pathology of IHPS, theories of aetiology, and associations. (see also Fluids/Nutrition/Growth)

**Chylous Ascites and Chylothorax:** The Trainee will describe the anatomy of the thoracic cage. The Trainee will discuss the etiologic factors which can lead to the formation of chylous ascites and chylothorax. The Trainee will describe the associated pathological changes which can occur in chylous collections.

**Congenital Diaphragmatic Hernia:** The Trainee will discuss fetal circulation, transition to post-natal circulation and how this process affects the management of the neonate with CDH. The Trainee will discuss the patho-physiological processes observed in newborns with CDH, and analyse the clinical implications of these. The Trainee will evaluate the theories around the patho-physiology and its management.

The Trainee will have an understanding of how fetal intervention may have a role in changing the pathogenesis of these complications.

The Trainee will be familiar with genetic conditions that cause disease in neonates. In order to discuss this topic the Trainee will need to understand concepts around:

- Genetic and environmental determinants and mechanisms of developmental/embryological defects

- Chromosomal associations

- Types of genetic mutations their effects and the implications for investigation

- Inheritance incl- Dominant, recessive, x-linked, isoparental disomy

- Mechanisms for genetic investigations. Including linkage, Next Generation sequencing and variations of whole genome, exome etc. FISH, PCR, Sanger sequencing, Western and Southern Blot, Micro-array

The Trainee will employ these concepts when discussing conditions of surgical significance (such as Hirschsprungs disease).

The Trainee will be able to recognise and categorise other congenital anomalies that may be present in their patients such as syndactyly/polydactyly and talipes.

The Trainee will demonstrate familiarity with conditions and syndromes that can be identified in the neonatal period and have implications for the diagnosis of or management of surgical disorders. These include (but are not limited to);

- Down Syndrome (Trisomy 21)

- Edwards Syndrome (trisomy 18)

- Epidermiolyis Bullae

- Turners Syndrome

- Congenital hypothyroidism

- Cystic Fibrosis

### 3: Fluids/Nutrition/Growth

The Trainee will have a detailed understanding of normal fluid and electrolyte metabolism in all age groups. The Trainee will evaluate the effects of illness and injury on normal homeostasis, and formulate appropriate management strategies in these situations.

The Trainee will have an understanding of the historical derivation of normal fluid and nutritional requirements, and demonstrate an up to date understanding of recent literature on the use of parenteral fluids in children.

The Trainee will understand the nutritional requirements in all age groups. The Trainee will describe normal growth and development and identify deviations from normal.

The Trainee will be able to discuss the issues related to general anaesthesia in infancy and childhood. The Trainee will discuss analgesic modalities and their use in infancy and childhood. (Also relevant for multiple other sections)

**Normal homeostasis:** The Trainee will be able to categorise normal reference values (and derivation from normal) for vital signs –such as blood pressure – and how these vary due to age of the patient. The Trainee will recognise when surgical disorders can affect these values –such as surgical causes for hypertension.

**Trauma/Shock:** The Trainee will understand the physiological effects of hypovolaemia. The Trainee will discuss the evaluation and management of shock. The Trainee will discuss the physiological derangements that occur immediately and over time in shock, both treated and untreated.

**Infantile Hypertrophic Pyloric Stenosis:** The Trainee will explain the physiological and metabolic derangement associated with IHPS. The Trainee will create a management plan to address these derangements. The Trainee will describe the consequences of incorrect management of this disorder.

**Tongue tie:** The Trainee will describe the normal anatomy of the tongue and lip, and identify normal variants and abnormalities. The Trainee will discuss the evidence around tongue/lip tie and its management.

**Gastro oesophageal reflux:** The Trainee will discuss the anatomical and physiological factors associated with competence and incompetence of the gastro-oesophageal junction. The Trainee will discuss the normal physiological function of the distal oesophagus and the abnormalities that are associated with disease states.

The Trainee will discuss the entity of gastro-oesophageal reflux disease, the natural history, common complications and the contributory pathological conditions. The Trainee will demonstrate an understanding of the diagnostic tests utilized in GORD and evaluate the relative strengths and weaknesses of these based on an understanding of the evidence for each.

**Oesophageal disorders:** The Trainee will describe the congenital and acquired causes of oesophageal stricture including the damage from caustic injury.

The Trainee will discuss the condition of achalasia; its etiology, presentation, pathology, investigation and natural history.

**Intestinal insufficiency:** The Trainee will demonstrate an understanding of normal bowel length (at all ages) and function. The Trainee will discuss intestinal insufficiency and demonstrate an understanding of its anatomical and physiological causes and consequences. The Trainee will describe in detail pathological processes that contribute to intestinal insufficiency. The Trainee will describe the metabolic and physiological consequences of intestinal insufficiency.

In addition to the embryological implications the Trainee will describe how the GI tract is affected functionally in gastroschisis, this should include an understanding of ante and perinatal complications, the effects of all forms of surgical management, and sepsis etc. that may be related. An understanding of abdominal compartment syndrome is essential here (and in other contexts eg. Trauma)

An understanding of the anatomic and metabolic disorders of the gut will be essential in addressing the learning aims related to other GI tract pathology in embryology, neonatal, trauma/burns, and acquired intestinal disorders.

**Liver**

The Trainee will be able to discuss pathological processes that cause liver failure in infancy and childhood. They will be able to discuss the specific histologic and pathologic process that occur in liver damage, normal healing, and pathological progression of liver disease.

The Trainee will outline the pathophysiologic consequences and the causative mechanisms of portal hypertension

**Cystic fibrosis and meconium ileus**

The Trainee will know the effects of cystic fibrosis on exocrine function especially related to respiratory, pancreatic and gut function. (See also neonatal) The Trainee will discuss the variety of ways Cystic Fibrosis can present outside of the neonatal period, and the ways known cystic fibrosis can present as or complicate management of surgical disorders.

**Obesity**

The Trainee will recognise how obesity complicates surgical management (both anatomical and physiological issues).

The Trainee must be familiar with bariatric surgical techniques and issues, especially the long term effects on physiology, growth, and nutrition.

**Constipation and faecal incontinence**

The Trainee will discuss the medical and surgical conditions that present in this manner. The Trainee will understand the physiology of normal bowel function and how deviations from normal contribute to clinical manifestations.

### 4: Trauma/Burns

In addition to the specific learning points below the Trainee will have an understanding of the anatomic, physiologic and metabolic derangements that occur in all traumatic injury. The level of knowledge required is commensurate with the Trainee's role in the acute management of trauma in childhood.

The Trainee will build on their knowledge of the normal inflammatory and coagulation cascade to discuss situations where disorder functioning will have implications for clinical management.

The Trainee will demonstrate detailed knowledge of how differences in paediatric anatomy, physiology, and behaviour contribute to different mechanisms, patterns of, and response to injury when compared to adults.

Successful completion of EMSB and EMST is assumed in these learning objectives.

The Trainee will demonstrate understanding of the socio-economic factors that influence risk for, presentation with and outcome from trauma and burns.

#### **Burns**

The Trainee will define Jackson's zones of injury, and describe the local and general effects of thermal injury.

The Trainee will demonstrate an understanding of the depth of injury after burning. The Trainee will discuss contributing factors, assessment of and sequelae of burns of differing depths.

The Trainee will describe the progression of burns of different depths. Identify epidemiology of paediatric burns trauma and apply prevention strategies and patterns of injury.

(In addition to Fluids/growth/nutrition) The Trainee will describe the circulatory

effects of burns trauma and the management of this.

The Trainee will identify the mechanism of circulatory impairment in a burnt limb, and the mechanism of impairment of respiratory excursion after trunk burns.

The Trainee will describe the process of scarring and scar contracture including scar maturation.

#### **Inhalation injury**

The Trainee will explain the mechanism of injury of the three types of inhalation injury (systemic intoxication, upper airway burns and broncho-pulmonary).

#### **Upper GI Burns**

The Trainee will describe the Pathophysiology of different caustic and chemical injuries of the mouth oesophagus and stomach including natural history of non treated chemical ingestions injuries.

**Trauma/Shock:** The Trainee will understand the physiological effects of hypovolaemia. The Trainee will discuss the evaluation and management of shock. The Trainee will discuss the physiological derangements that occur immediately and over time in shock, both treated and untreated.

The Trainee will demonstrate understanding of resuscitation in terms of both blood volume and blood components. This will include knowledge of components and adjuncts to resuscitation and investigations and monitoring of successful resuscitation.

The Trainee will have an understanding of how products and devices can be used intra-operatively to control bleeding, in particular the strengths and limitations of commonly used products.

The Trainee will describe the systemic physiological effects of crush injury to a limb.



### **Abdominal trauma**

The Trainee will describe abdominal anatomy in detail, describe the types of injury to hollow and solid abdominal viscera, and the abdominal wall, in blunt and penetrating trauma.

The Trainee will discuss how patterns of injury in childhood relate to specific behavioral, anatomical and physiological characteristics.

The Trainee will discuss the evidence that supports non-operative management of paediatric solid organ injuries.

The Trainee will discuss the pathophysiologic sequelae of injury to abdominal organs with reference to presentation (including delayed presentation) natural history and long term outcomes.

### **Neck Trauma**

The Trainee will be able to discuss anatomical considerations in relation to neck trauma and patterns of injury.

### **Chest trauma**

The Trainee will describe thoracic anatomy, relating internal viscera with surface anatomy. The Trainee will describe the mechanisms of injury that produce haemothorax and pneumothorax, and their physiological consequences.

The Trainee will discuss the additional issues surrounding tension pneumothorax.

### **Spontaneous pneumothorax**

The Trainee will understand the pathophysiology of spontaneous pneumothorax and how this influences investigation and surgical treatment.

The Trainee will list risk factors for and conditions associated with spontaneous pneumothorax.

### **Other Chest Trauma**

The Trainee will describe the patho-physiology of cardiac tamponade and its complications.

The Trainee will describe mediastinal injuries and their mechanisms.

The Trainee will identify lung injuries and their consequences in addition to haemo/pneumothorax.

### **Soft tissue trauma**

The Trainee will recognise the types of injury that are associated with different mechanisms of trauma.

The Trainee will recognise the likely bacteriological factors in different mechanisms of trauma. Describe the pathophysiology of aerobic and anaerobic infection of soft tissue following trauma.

The Trainee will be able to discuss the pathology and physiology of bone injury and healing, consistent with the knowledge obtained for the GSSE.

## **5: Infection/inflammation**

The Trainee will build on their GSSE knowledge and demonstrate an understanding of the physiological processes involved in inflammation and infection (see also Trauma/Burns). They will understand how inappropriate and ineffective inflammation and healing can have implications for surgical patients, and produce disease states that may require surgical management.

The Trainee will understand how effective inflammation is essential for surgical healing.

### **Systemic**

The Trainee will discuss the processes involved in the clinical picture of SIRS/MODS/CARS1 including the role of cytokines.

The Trainee will understand when other conditions or treatments can affect these processes e.g. immune suppression.

The Trainee will discuss the role of vaccination in childhood in particular their effects on surgical pathology. Examples Hib and streptococci vaccines both in relation to the alteration on surgical presentations and their importance in relation to splenectomy. The Trainee will be familiar with OPSI.

### **Cervical lymphatic node infections**

The Trainee will discuss the epidemiology and pathogenesis of cervical lymphatic infections. In particular Lymphadenitis; Acute abscess; Subacute/chronic lymphadenitis; MAIS complex

### **Salivary gland pathology**

The Trainee will discuss epidemiology and pathogenesis of disorders of the salivary glands including Infections; Inflammatory disease; Calculi; Tumour; Ranulas/mucoceles

### **Empyema Thoracis**

The Trainee will list the causative organisms for empyema

The Trainee will describe the patho- physiological sequence of events in the formation of empyema, and the long term complications associated with this disease process.

### **Bronchiectasis**

The Trainee will understand the pathophysiology of bronchiectasis and the types described. The Trainee will describe the causes, possible associations and the microbiology of the condition.

### **Appendicitis**

The Trainee will have a thorough knowledge of the differing pathological entities that can affect the appendix and how these can result in a variety of patho- physiological states.

### **IBD**

The Trainee will know the key gross and histological pathological findings, complications and the aetiological theories

### **Perianal disease**

The Trainee will describe the different pathologies, and their etiologies, that can present within the perianal region in childhood

### **Skin and Soft tissue infection**

The Trainee will explain the natural history of HPV and molluscum contagiosum skin infections.

The Trainee will explain the pathophysiology of cellulitis and necrotising fasciitis. They will identify risk factors and discuss disease progression and complications.

### **Hydatid Disease**

The Trainee will describe the life cycle of Echinococcus. The Trainee will appreciate how this disease is eradicated in New Zealand. The Trainee will describe the process by which Echinococcus causes disease in humans. The Trainee will identify the complications of Echinococcal disease both untreated and related to treatment.

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<sup>1</sup> Systemic Inflammatory response Syndrome/Multi-Organ Dysfunction Syndrome/Compensating Anti-inflammatory-Immune System

**Tuberculosis**

The Trainee will describe the development of Tb infection in humans including concepts of primary and disseminated infection, and latent and active disease. The Trainee will be able to identify situations in which Tb may present as or mimic surgical disease.

**Surgical considerations**

The Trainee will demonstrate understanding of how surgical trauma affects the patient.

In particular the effects of laparoscopy/thoracoscopy on normal physiology and its pro-inflammatory effects.

Effects of surgery on temperature homeostasis and fluid management

The effects of surgical devices on tissues such as diathermy and advanced co-agulative/dissecting devices

**Systemic conditions with surgical implications**

The Trainee will have knowledge of systemic illnesses and processus that affect or mimic surgical disease or its treatment. For example Henoch Schonlin Purpura, Haemolytic Uremic Syndrome, Graft v Host disease etc.

### 6: Neoplasia

The Trainee will demonstrate familiarity with genetic and biochemical processes and techniques that are utilized in the research into and management of paediatric tumours.

The Trainee will demonstrate detailed knowledge of paediatric solid tumours that are managed by paediatric surgeons. The Trainee will also have some familiarity of the oncological conditions that are commonly treated in childhood and have implications for the practicing paediatric surgeon.

#### **General oncologic considerations**

The Trainee will demonstrate a familiarity with chemotherapeutic agents, their mechanisms of actions and subsequently they side effects that will have implications for surgical patients. (e.g. tumour lysis, typhlitis, thrombocytopenia etc.)

The Trainee will demonstrate familiarity with radiotherapy techniques, their side effects and surgical issues relating to the use of radiotherapy, particularly its use as an adjunct or alternative to surgical management of specific tumours. The Trainee will be familiar with nuclear medicine techniques of diagnosis and treatment in relationship to paediatric oncology.

The Trainee will describe hereditary cancer syndromes of relevance to paediatric practice.

The Trainee will demonstrate an understanding of genetic mutations and translocations that are of diagnostic, prognostic or therapeutic importance i.e. EWS/FLY Ewings or FOX/PAX translocations for Rhabdomyosarcoma.

The Trainee will be familiar with BMT and its effects related to potential surgical intervention, incl GVHD.

#### **Vascular Anomalies (See also Neonatal and Head & Neck Modules)**

The Trainee will explain the commonly accepted classification of vascular malformations, and describe the advantages of this system over previous nomenclatures.

The Trainee will discuss the current theories of pathogenesis of vascular anomalies, their development and natural history.

The Trainee will describe the embryology of lymphatic system related to sites of appearance of lymphangioma.

The Trainee will describe the vascular syndromes seen in the neonatal period

The Trainee will discuss the long term issues related to vascular malformations throughout childhood and into adulthood.

#### **Soft tissue tumours and Rhabdomyosarcoma (See also Skin Module)**

The Trainee will describe the histological subtypes of rhabdomyosarcoma and their prognostic implications

The Trainee will recognise the different tumour types in specific sites (e.g. pelvic, head & neck) and for different age groups and relate to clinical prognosis and tumour behavior.

The Trainee will list a classification of soft tissue tumours of childhood including PNET and fibromatosis groups

#### **Neuroblastoma**

The Trainee will demonstrate a thorough understanding of neuroblastoma.

The Trainee will explain the origin of neuroblastoma and the potential sites of their manifestation and describe the genetics and histological features and their prognostic implications.

The Trainee will demonstrate knowledge of the metabolic tumour by-products and relate this to clinical presentations and their consequences.

The Trainee will understand the prognostic framework employed for staging neuroblastoma. This will include detailed understanding of genetic, cytologic and clinical staging criteria.

The Trainee will discuss the maturation process seen in neuroblastomas (this should include understanding of the untreated natural history in addition to treatment effects).

**Nephroblastoma (Wilms tumour) & Mesoblastic nephroma**

The Trainee will describe the pathogenesis of Wilms tumour and its histological subtypes.

The Trainee will identify the genetic mutations in and the various syndromes associated with the development of Wilms tumour and be able to discuss prognosis and screening in at risk individuals.

Describe the prognostic implications of nephrogenic rests in patients with and without Wilms tumours.

Identify the pathological features of mesoblastic nephroma and its prognostic implications

The Trainee will discuss the condition of congenital cystic mesoblastic nephroma and its similarities and differences to Wilms tumour.

The Trainee will demonstrate knowledge of other renal tumours that can occur in childhood, and discuss their differences to nephroblastoma.

**Gonadal tumours**

The Trainee will describe the classification of tumours of the gonads with respect to the cell lines of origin of these tumours.

The Trainee will demonstrate a detailed understanding of germ cell tumours of the testis and ovary and how they relate to and differ from teratomas in other sites.

The Trainee will list other benign and malignant gonadal tumours. Discuss their presentation and diagnosis in relationship to their histologic type and cell function. The Trainee will describe natural history and prognosis in these tumours.

The Trainee will discuss how dysplasia in gonads is associated with tumour development and relate this to relevant clinical situations.

**Lymphoma**

The Trainee will describe the histological classification of lymphomas.

The Trainee will identify clinical situations in which lymphoma can present as or complicate general surgical/urological diseases.

The Trainee will discuss the staging of lymphoma.

**Teratoma/ Sacrococcygeal teratoma**

The Trainee will explain the embryologic origin of teratoma and list the sites where teratoma may develop.

The Trainee will describe the histological features of mature, immature and malignant teratoma.

The Trainee will discuss tumour markers and their role in diagnosis, prognosis, and monitoring for teratomas.

**Hepatic & Pancreatic tumours**

The Trainee will list the primary tumours arising from the liver and pancreas

The Trainee will describe the histological features of hepatoblastoma and hepatocellular carcinoma

The Trainee will classify the tumours of the pancreas

**Hamartomas (excluding congenital vascular anomalies)**

The Trainee will define a hamartoma and describe the histological features of various types of hamartomas (e.g. hamartoma of infancy, mesenchymal hamartoma).

**Intrathoracic masses**

The Trainee will list the causes of intra thoracic masses in children. The Trainee will describe the patho-physiological consequences of intra-thoracic masses and their implications, including peri-operative concerns.

**Thyroid & Parathyroid**

Nodules; Malignancy; Thyroiditis; Goitre; Ectopia

The Trainee will discuss thyroid and parathyroid pathologies presenting in childhood and adolescence.

The Trainee will describe aetiology and familial factors in paediatric thyroid disease especially with reference to M.E.N

### 7: Other Acquired abdominal disorders

The Trainee will demonstrate their knowledge of the epidemiology, presentation, pathogenesis investigation and natural history of disorders of the abdomen that present as disease states in paediatric surgical practice. This knowledge will be of the level of detail to allow discussion of these conditions in terms of normal presentation and diagnosis. This knowledge will allow the Senior-SET Trainee to develop their clinical practice to the level required to complete the SET program.

(Related to discussions in almost all other sections) The Trainee will demonstrate and understanding of radiologic and nuclear medicine techniques as they pertain to the management of paediatric patients. The Trainee will understand the technical issues around how x-ray, CT, MRI, Ultrasound, relevant Tc99 radiolabeled techniques (DMSA, MAG3, DTPA, HIDA, DISIDA, MIBG, etc.) and PET scanning are used in paediatric surgical practice, their strengths and limitations. The Trainee will be able to discuss the morbidity associated with clinical imaging most importantly the issues around exposure to ionizing radiation.

#### **GI bleeding**

The Trainee will develop a framework to discuss the potential sites of GI bleeding in children and relate this to the common pathologies seen in children.

#### **Gastrointestinal polyps**

The Trainee will understand the pathology of polyps of the intestine in children including any relevant genetic disorders.

#### **Splenic disease including trauma**

The Trainee will describe the pathological conditions that can affect the spleen, the integral function of the spleen and the consequences of its absence.

The Trainee will use their knowledge of normal and disordered physiology to describe the importance of the Early Management of Severe Trauma (EMST) principles.

#### **Hepatobiliary disease**

The Trainee will know the pathology and pathophysiology of the different hepatobiliary conditions that present to paediatric surgeons including aetiological theories where relevant.

(Biliary Atresia see Neonatal)

#### **Pancreatic disease**

The Trainee will outline the embryology of the foregut and the function of the pancreas gland; discuss the pathophysiology of pancreatitis and endocrine dysfunction of the pancreas in disease processes relevant to childhood including presentation MET in children.

The Trainee will discuss anatomical variations of the pancreas that contribute to disease states.

#### **Choledochal Cyst**

The Trainee will describe the anatomy of the biliary and portal system, know the classification for choledochal cysts and have an understanding of the pathologic changes in the biliary and pancreatic system which may contribute to the development of this abnormality. The Trainee will discuss and compare the theories of etiology of choledochal cysts. The Trainee will describe the long term issues related to choledochal disease.

#### **Abdominal cysts**

The Trainee will discuss the different abdominal structures which may give rise to an intra-abdominal cyst. The Trainee will understand the anatomy of those structures and the pathological changes associated with the cyst.

**Rectal Prolapse**

The Trainee will describe the process of prolapse in children and its relationship to causative etiologies including Cystic fibrosis, constipation and connective tissue (CT) disorders.

The Trainee will discuss normal sphincter and normal submucosal development in children.

**Intussusception**

The Trainee will describe the pathology of intussusception and the complications. This should be related to the presentation and common clinical features.

The Trainee will explain the idiopathic and pathological lead points that cause this condition, relating them to the age of presentation and the physiological derangements.

**Recurrent abdominal pain of childhood**

The Trainee will outline the clinical features of this condition including differential diagnosis



**8: Genito-Urinary**

**Urinary incontinence**

The Trainee will describe the embryology and anatomy of the urogenital tract and the normal development of urinary continence

**Neuropathic / neurogenic bladder**

The Trainee will describe the anatomy and innervation of the bladder and sphincter and list the causes of neurogenic bladder

The Trainee will classify neurogenic bladder with reference to compliance and leak point pressure, and subsequent outcomes such as continence, infection and potential for renal damage (uropathy).

**Vesicoureteric reflux and UTI**

The Trainee will describe the embryology and anatomy of the urinary tract related to the condition of vesicoureteric reflux and the associated findings. Discuss theories of natural progression.

The Trainee will discuss how abnormalities and dysfunction of the renal tract contribute to UTI and how UTI contributes to dysfunction of the urinary tract.

**Ovarian Torsion**

The Trainee will discuss predisposing conditions for ovarian torsion, discuss ovarian torsion in normal ovary and classify ovarian cysts.

**9: Skin/Subcut/bodywall/extremities**

**Naevus**

The Trainee will discuss the types of naevus affecting children. They will describe the maturation of normal nevi. The Trainee will describe the pathological features of abnormal nevi, in particular describe Spitz naevus and discuss its clinical implications.

**Benign soft tissue lesions.** (See also Childhood Tumour Module)

The Trainee will describe the pathology of pilomatrixoma, juvenile xanthogranuloma, granuloma annulare, nodular fasciitis and fibrous hamartoma of infancy

**Breast swelling**

The Trainee will describe the embryology of the breast and hormonal effects on it.

**Gynaecomastia**

The Trainee will describe the embryology of the breast and its normal and abnormal development in males. The Trainee will develop a list of potential aetiological factors.

**Disordered Toenails**

The Trainee will explain the pathology of disordered toenails and their complications in infancy and adolescence, describe how they differ.

**Palmar hyperhydrosis**

The Trainee will discuss hyperhydrosis and its cause, and use this knowledge to understand its treatment.

**Hydradinitis Suppurativa**

The Trainee will discuss the pathology and natural history of hydradinitis Suppurativa.

**Disorders of growth:**

The Trainee will describe disorders of growth and their implications for surgical disease. For example hemi-hypertrophy.

The Trainee will identify scoliosis and describe the types.

**Chest wall deformity**

The Trainee will describe chest wall deformities and understand their anatomic and physiological consequences

**Pilonidal Sinus**

The Trainee will recognise the pathology of pilonidal sinus in the natal cleft and other locations. The Trainee will describe the pathological process of pilonidal sinus formation, its pathologic progression and its long term outcomes.