Year 2 Peer Based Learning 2020

Miscellaneous Answers

Please note – this learning resource has been produced by the GUMS Academic Team. It is possible that there are some minor errors in the questions/answers, and other possible answers that are not included below. Make sure to check with other resources.

Case 1

You are the intern on the paediatrics team, and you are sent to do a routine baby check on Adam, a 2 year old baby. The parents have called you to do this check, after the UQ student did one but couldn't take a history from the parents. You, as the supreme historian, find out that Adam hasn't passed a bowel motion yet.

What are the differentials for this presentation?

- *Hirschprung's disease*
- *Meconium ileus - usually from cystic fibrosis*
- *Meconium plug syndrome*
- *Atretic disease e.g. anal atresia*

How are most cases of cystic fibrosis diagnosed in Australia? How is CF tested for? What else is tested for in this screening program?

- *Newborn screening program - heel prick blood test*
- *CF: detects 'immunoreactive trypsinogen' - the basis of this test is that there is impaired release of pancreatic enzymes in CF. this 'immunoreactive trypsinogen' has not been converted to its active form, and is hence abundant in the blood of patients with CF*
- *Screening program also tests for congenital hypothyroidism, galactosemia and phenylketonuria*

When Adam becomes 5 years old, is he more likely to have features of malabsorption or respiratory infection?

- Malabsorption - GI symptoms are common in infancy
- Respiratory symptoms are common in adolescence and adulthood!
- (but they will get both early of course)

What is the most common mutation in CF?

- dF508 - deletion in the amino acid in the 508th position of the Cl- channel protein

In terms of pathophysiology of CF, what is the direction that the Cl- channels pump Cl- in the sweat glands, pancreas and lungs? What is the significance of this?

- In sweat glands, the channel REABSORBS Cl-
  - Failure to reabsorb Cl- = salty sweat
- In the lungs and pancreas, the channel EXCRETES Cl-
  - Less Cl- excretion → Na+ is not pumped out with the Cl- → retained NaCl draws in water to cells → **hyperviscous secretions due to water depletion**

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Complete the following table on clinical features of organ systems affected in CF

THE HIGH YIELD ONES ARE IN BOLD

GIT	**- meconeum ileus** **- FTT (failure to thrive)** **- pancreatic disease: steatorrhoea, malabsorption, DM etc.** - liver and bile duct - cholecystitis, cholestasis - intestinal obstruction
Resp	**- bronchiectasis with chronic productive cough, wheeze and recurrent pulmonary infections** **- clubbing (related to chronic hypoxaemia)** - chronic sinusitis, polyps - ABPA
MSK	- osteopenia - kyphoscoliosis
Sweat glands	**- salty sweat w/ electrolyte wasting**
Urogenital	- nephrolithiasis, nephrocalcinosis, UTIs **- men: obstructive azoospermia, absent vas deferens** **- women: viscous cervical mucous, amenorrhoea**

Disease of which of the above organ systems leads to mortality in CF patients?

- respiratory

At 7 years of age, Adam develops a respiratory infection, coughing copious amounts of productive sputum with a loud expiratory wheeze heard on auscultation. What is the most likely causal organism? What would the most likely organism be if he was 18?

- In childhood/INITIAL respiratory infections include *S. aureus* and *H. influenzae*
- During adolescence and adulthood, the main organism is *P. aeruginosa*

In relation to *P. aeruginosa* infection, why is early antibiotic treatment vital?

- Early on, antibiotics will ERADICATE the bacteria
- However, overtime, pseudomonas establishes a biofilm - a layer of 'slime' → antibiotic resistance occurs

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- Therefore, antibiotic resistance will allow for SUPPRESSION of an infection, but not ever eradicate the bacteria!
- Therefore, TREAT EARLY TO PREVENT BIOFILM FORMATION

Why does P. aeruginosa establish biofilms easily in CF?

- The mucous environment is ideal for biofilm formation

On spirometry, what pattern will be seen? What will happen to the total lung capacity and functional residual capacity?

- Obstructive - FEV1:FVC
- Both will increase due to air trapping

Define bronchiectasis. Why do CF patients get it?

- Irreversible and abnormal dilation in the bronchial tree that is generally caused by cycles of bronchial inflammation in addition to mucous plugging and progressive airway destruction
- The recurrent infections and mucous plugging leads to abnormal dilation

Why is mucous clearance impaired in CF?

- Mucociliary escalator impairment
- SUMMARISE TO THE STUDENTS: 1. Hyperviscous mucous secretions 2. Impaired mucous clearance from mucociliary escalator damage 3. Recurrent infections and bronchiectasis = REASONS FOR RESPIRATORY DAMAGE

At 22 years of age, Adam develops acute onset epigastric pain that radiates to the back. He does not want to eat at all and has vomited twice now. What is the most likely diagnosis? What investigations do you want to do straight away?

- Acute pancreatitis
- FBC - see if they are actively bleeding, look for leukocytosis
- LIPASE = confirms diagnosis *(bonus info: it's diagnostic, not prognostic. I.e., high lipase tells you that there is pancreatitis. The trend of lipase or down doesn't reflect the severity of the disease)*
    - Amylase is rubbish compared to lipase (low specificity)
- Serum calcium
- Main one they should say is lipase
- Etc. etc.

What is the reason Adam developed this? In patients who don't have CF, what are the two most common causes of this pathology and what investigations/history would help you establish the aetiology here?

- Mucous plugging in pancreatic duct → enzymes have trouble reaching duodenum → with a longer time it takes to go to the duodenum, there is more chance of premature activation of the

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enzyme precursors (zymogens) → enzymes activated within the pancreas instead of the duodenum digest the pancreas - this is pancreatitis

- Most common aetiologies are gall stones (choledocholithiasis) and alcohol
    - For gallstones - abdo US is good to see the stones, and LFT is good to see high ALP, GGT and bilirubin
    - For alcohol, the history is the key - recent binge

What are the cornerstones of management for the respiratory and GI complications of CF? Break it up into lifestyle, medical and allied health

- RESP
    - Lifestyle = avoid sick contacts, clear mucous frequently
    - Mucolytics: dornase alfa, hypertonic saline and inhaled mannitol
    - Bronchodilators only used if co-existing asthma or evidence in big increase in FEV1 following administration
    - Antibiotic therapy - vancomycin for S. aureus in early life, tobramycin plus others for P. aeruginosa in adulthood - don't learn specific antibiotics, just a bonus
    - Long term, ventilation and/or lung transplant is needed
    - Main allied health is CHEST PHYSIO - to do mucous clearance exercises
- GI
    - High calorie diet
    - Give pancreatic enzymes and substitute fat soluble vitamins (ADEK)
    - Main allied health is DIETICIAN - give counselling on appropriate foods to eat
- Other allied health who are important - psychology - multifactorial condition with mental health issues a common feature - important to think about in these patients!

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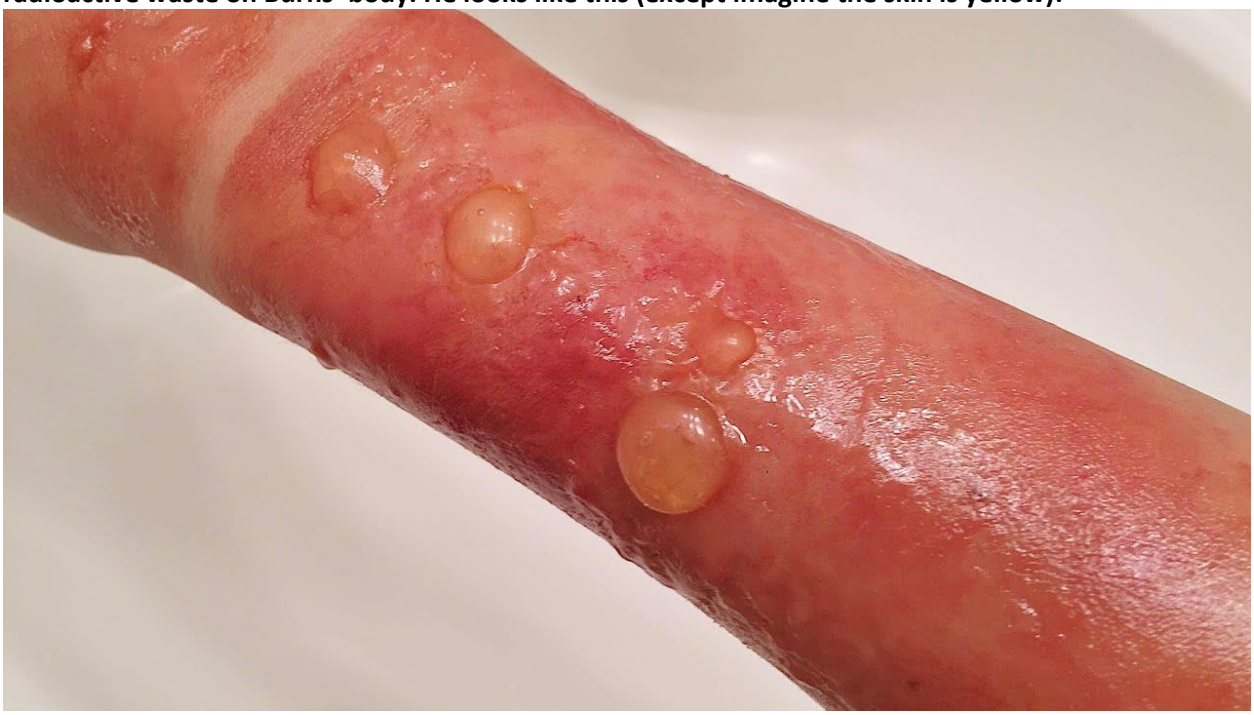
Case 2

Montgomery Burns is brought in by his assistant and sidekick, Smithers, into Springfield hospital following a burn. They were plotting to irradiate Springfield, when Smithers accidentally poured radioactive waste on Burns' body. He looks like this (except imagine the skin is yellow):



Mr Burns has a superficial partial thickness (2nd degree) burn, as depicted above. Why is this a superficial partial burn?

- Erythematous
- Swelling
- **Presence of vesicles/bullae** does **NOT** occur in 1st or 3rd degree burns

If on examination, this burn was non-blanching, and the patient was in extreme pain, what exact type of burn would they most likely have?

- Deep dermal (deep 2nd degree) - there are involvement of the nerve fibres (minimal pain) and blood vessels (minimal blanching)
- So, say to the students: you need to look at the burn, examine it for blanching and ask for pain to confidently assess the type of burn

If Smithers called you when they were back at the nuclear plant before coming into hospital, what advice would you give in regards to first aid for Mr Burns?

- Run under cool water for 20 minutes - NO ICE

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- Apply cling wrap after cooling to limit heat loss and evaporation of fluids
- ABCs - assess airways, breathing and circulation - ESPECIALLY in inhalation burns!
- Provide analgesia
- Remove burnt clothing

When Mr Burns comes in, your boss, Dr Nick (he is waving to you below), asks you to calculate the total surface area of his burn (+/- 2% is fine). He has second degree burns over his entire back, excluding his buttocks, the anterior aspect of his R arm and hand, and all of his R leg at and below the knee. The anterior aspect of his L arm has a first degree burn over it.

- Back without buttocks (buttocks included with legs!) = 18%
- R leg below knee (front and back) = 9%
- Anterior aspect of R arm and hand = 4.5%
- First degree burns don't count! So don't count L arm!
- Therefore, approximately 31.5%

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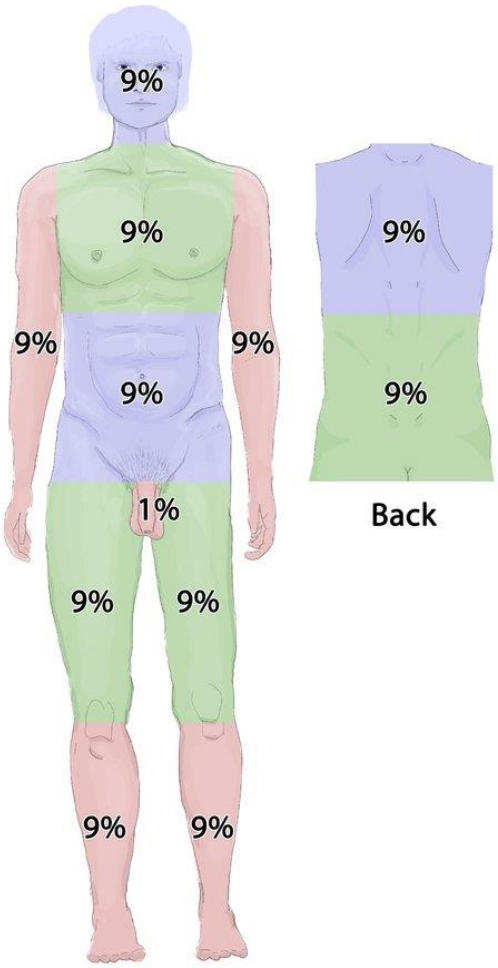

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What formula is used to govern fluid therapy? What type of fluid is given? When is the fluid given?

- **Parkland formula: 4 mL x %TBSA x weight (kg)**
- Give a crystalloid e.g. saline, not a colloid
- First half in the first 8 hours, second half in the next 16 hours
- This is to prevent complications of over resuscitation e.g. pleural effusions
- This formula DOES NOT account for maintenance fluids - this only accounts for fluids lost in the burn. Therefore, if they can't tolerate oral fluids, you have to ALSO give maintenance fluids!

Fill in the following table regarding clinical features of the different types of burns

	1st degree - superficial	2nd degree - superficial dermal	2nd degree - deep dermal	3rd degree- deep/full thickness
extent	epidermis with intact epidermal barrier	epidermis and papillary (upper) layer of dermis	epidermis and further into dermis	all three
cause	classically sunburn	chemical, thermal etc.	chemical, thermal etc.	chemical, thermal etc.
signs and symptoms	painful, dry, red, warm, blanching, erythematous	most painful, moist, red, weeping, blisters, warm, blanching, erythematous	no blanching, minimal to no pain, blisters, moist pale or red (depending on extent)	no blanching, painless, waxy white/leathery gray/charred black dry and inelastic
healing	heals spontaneously	spontaneously re-epithelialise from dermal remnants (more common); time depends on depth - longer it takes, more chance of hypertrophic scar	same as left box	always heal by secondary intention —> scar
other notes	not included in assessment of TSBA burnt			can go into muscle and bone (esp. if electrical) —> cause things like rhabdomyolysis

Mr Burn's labs show a high creatinine and high K+. What is the most likely diagnosis?

- AKI secondary to hypovolaemic shock

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Apart from the above, list broadly the other complications to think about in burns patients

- Shock → end organ damage
- Compartment syndrome
- Acute respiratory distress syndrome (ARDS) (poor prognosis)
- Severe laryngeal oedema in inhalation burns
- Long term scarring - consider psychological impact on patient etc.

Since the entire circumference of Mr Burn's R leg was involved, what surgical procedure should be done and why?

- Escharatomy
- For circumferential burns, escharotomy lowers the risk of compartment syndrome

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Case 3

Dorothy has presented to her GP twice previously in the last 1.5 months with recurrent URTI like symptoms. This UQ GP failed to take a thorough history and kept dismissing this as colds. You as the knowledgeable Griffith graduate dig deeper and find that Dorothy has multiple sexual partners and has previously had pelvic inflammatory disease.

In a patient with recurrent colds and a previous history of STIs, what diagnosis must be tested for?

- HIV

How is HIV transmitted?

- Sexual, parenteral, perinatal

What type of virus is HIV? Briefly discuss some of the molecules involved in its replication cycle

- Virus gp120 molecule binds CD4, as well as a coreceptor, either CCR5 on macrophages (early infection) or CXCR4 on T cells (late infection)
- injects single stranded RNA into cell
- uses reverse transcriptase to make DNA
- uses integrase to combine with host DNA
- host DNA transcribes DNA —> makes viral mRNA which is translated —> viral protein produced
- cleaved with protease inhibitors to form functional bits of viral protein —> buds out, taking host cell membrane with it —> forms new virus

List the other common sexually transmitted infections

- Chlamydia
- Gonorrhoea
- Syphillis
- HPV – this is unique compared to other STIs because it is transmitted in spite of use of condoms.
- HBV
- HCV

What is meant by the 'window period'? What is the significance of this when testing for HIV?

- it takes **3 months for an individual to produce enough Abs to be detected by the tests - so, if suspect HIV, ask p to return in 3 months for another to test to confirm if they have HIV or not**

With making reference to the below graph, why can patients remain asymptomatic for many years following initial repeated bouts of flu like illnesses?

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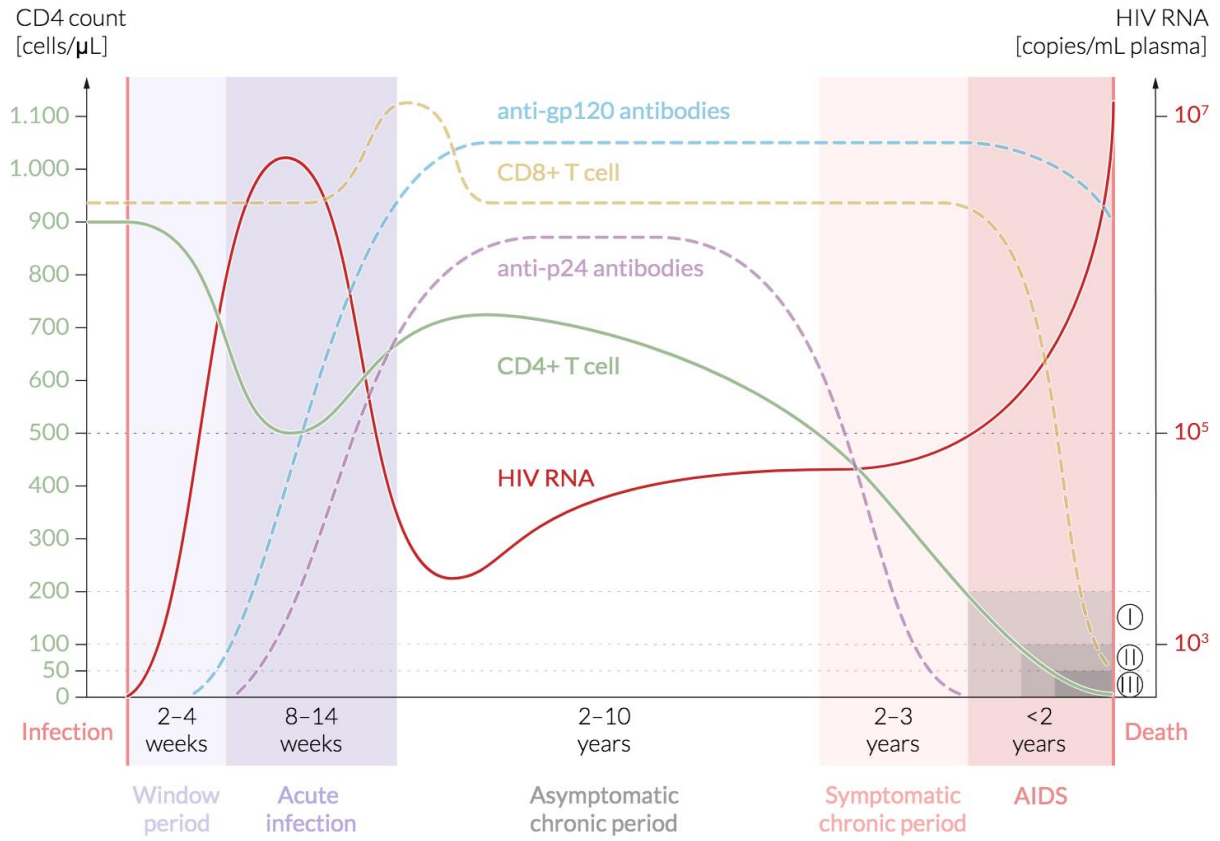

- Following acute infection, there is a latency period
- This is where immunity is built to the virus, which causes a drop in HIV viral load and increase in CD4 T cell count
- However, over time, the CD4 T cells die off and the viral load increases

What kind of infections will the patient get when their CD4 T cell count is <500? How about <200? What is the most common cause of death in patients with AIDS and what is the prophylaxis for this?

- Continuing on from above, <500 means they are now out of the latency period and are starting to have recurrent infections - these include oral candidiasis, generalised lymphadenopathy
- <200 is defined as AIDS. here they will get reactivation of TB, oesophageal candidiasis, Kaposi sarcoma etc. etc. etc.
- The most common cause of death in AIDS patients is **PCP - Pneumocystic jirovecii.** AIDS patients receive bactrim (TMP-SMX) prophylaxis for this! This causes an atypical pneumonia with 'ground glass' appearance on x-ray

What is HAART? List the key anti-retroviral classes and state their MoA

- Note that there is a lot of detail here - they should judge how much they should learn based on the past exams
- NRTIs - nucleotide/nucleoside reverse transcriptase inhibitors e.g. tenofovir

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    - step in life cycle: replication of viral DNA
    - incorporate into proviral DNA chain —> competitively inhibit nucleotide binding to reverse transcriptase (they are structurally similar enough so that they can bind the DNA chain, but are different enough that they can terminate the enzyme)
- NNRTIs
    - step in life cycle: replication of viral DNA
    - bind to reverse transcriptase at a site different to NRTIs - causes conformational change of the enzyme —> reduces its activity
- protease inhibitors
    - step in life cycle: maturation of viral proteins
    - assembly of vision depends on on HIV-1 protease which cleaves the polypeptide products of HIV mRNA into their functional parts
    - therefore, protease inhibitors prevent maturation of new viruses
- integrase inhibitors
    - step in life cycle: integrate viral DNA into host DNA
    - Inhibits HIV genome integration into host cell via reversible inhibition of integrase
- fusion inhibitors
    - step in life cycle: attachment and entry of HIV into CD4 T cell