Cross Canada Rounds
Short Cases

Ajay Kevat MBBS (Hons), MMed, MPH, FRACP

Clinical Fellow: BC Children's Hospital

Research Fellow: UBC, Monash Children's Hospital & Murdoch Children's Research Institute (Royal Children's Hospital Melbourne)

Case 1

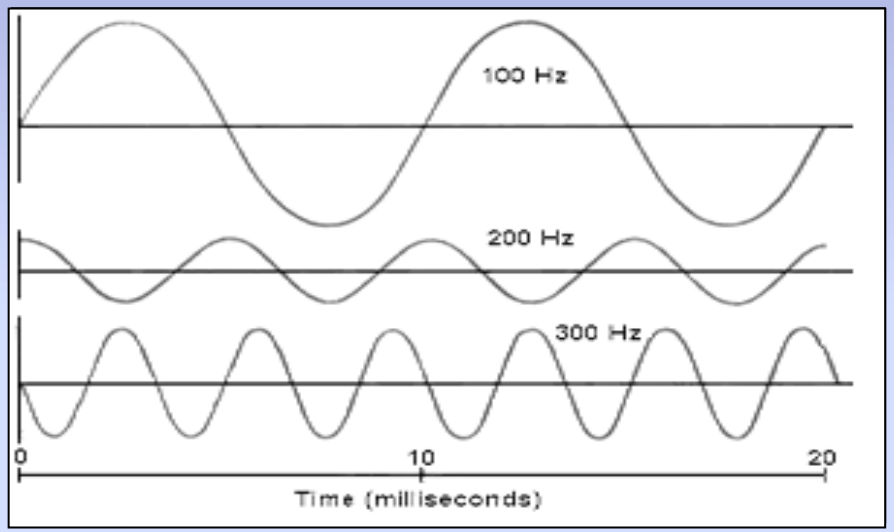
History

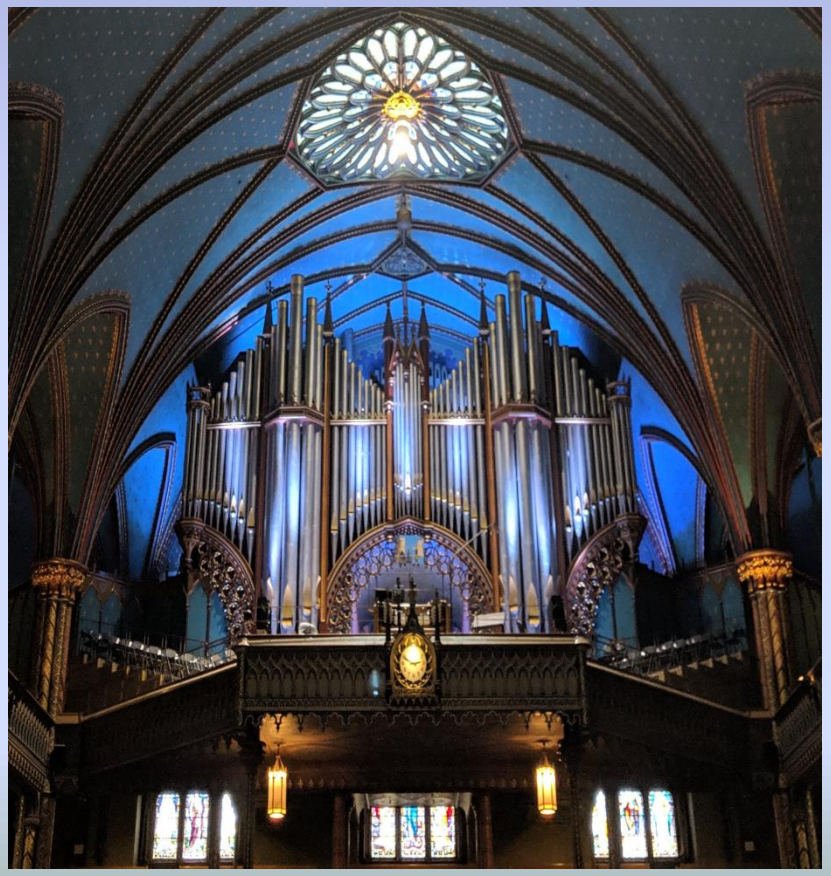
- 18 month old boy
- Born at term in Canada to non-consanguinous parents
- maternal active HPV lesions at time of delivery
- No neonatal resuscitation / admission required

- Bronchiolitis (11 months of age)
- URTI, with viral wheeze (16 mths), with initial response to po steroid & salbutamol but then had a short fever and return of wheeze

Case 1

What is wheeze?

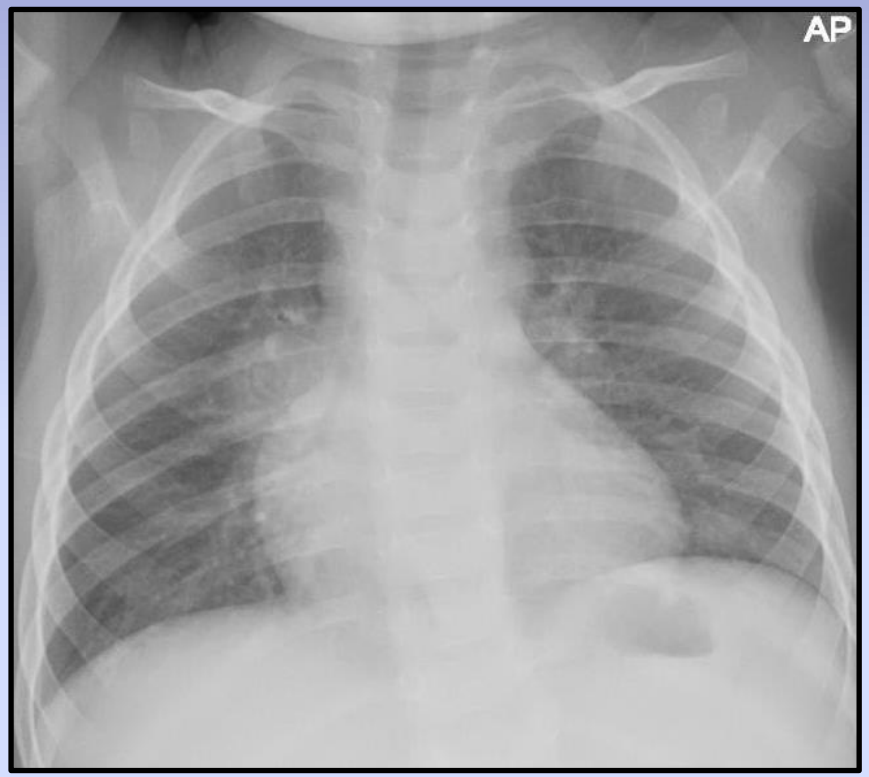




Case 1

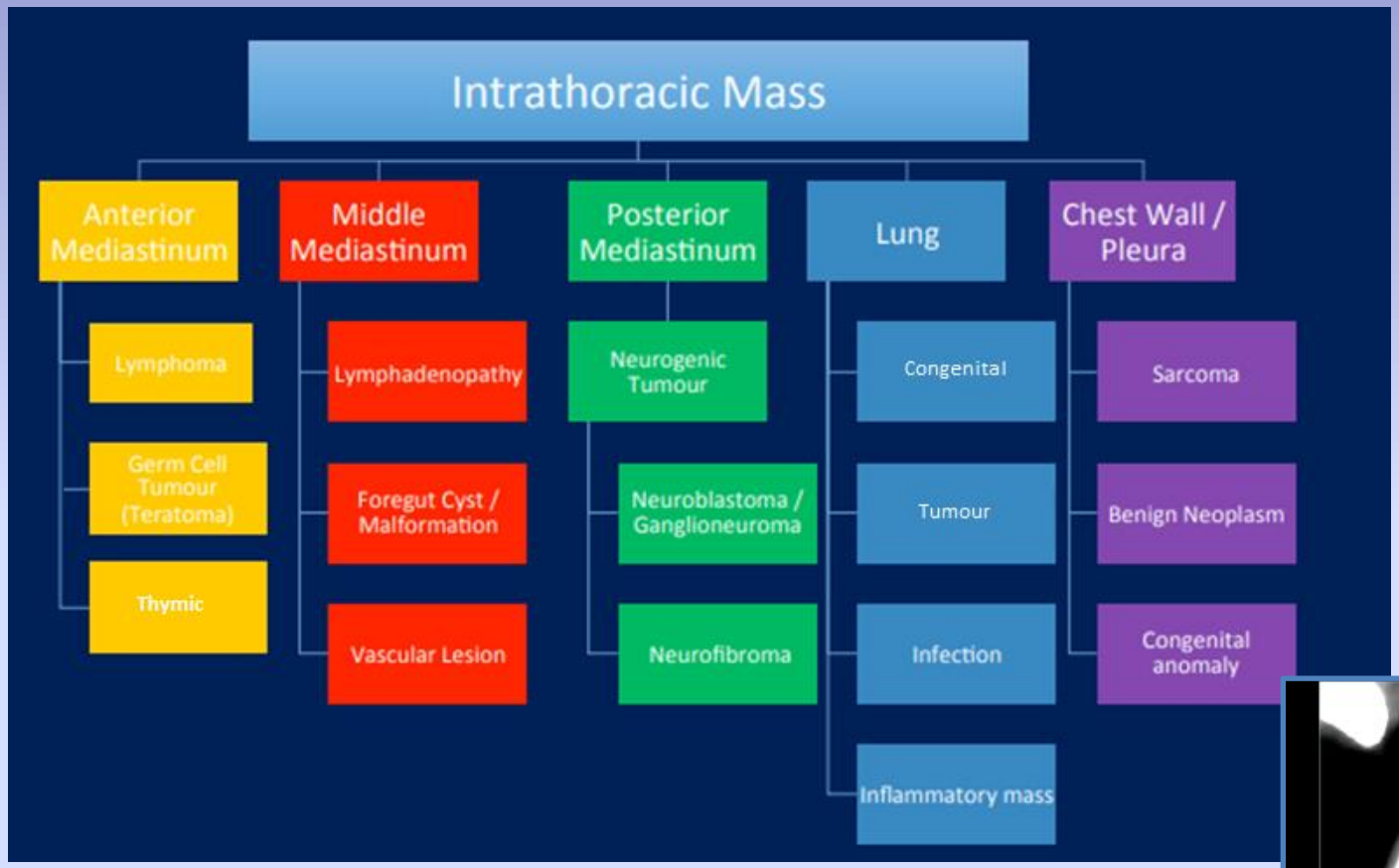
History

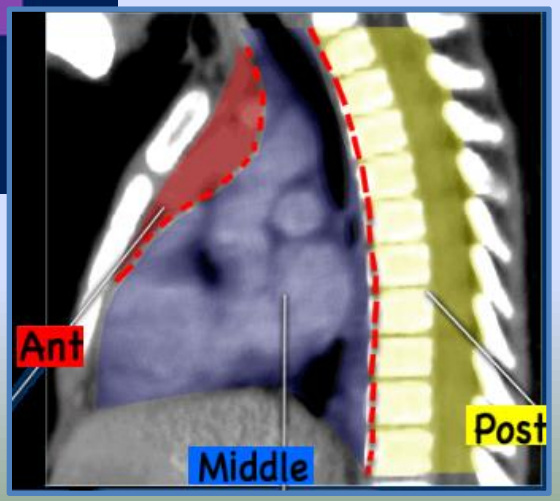
- CXR: 3cm diameter round pneumonia in superior segment of right lower lobe, so treated with IV abx; clinically improved → discharged
- Referred to resp. clinic due to persisting abnormality on XR



- No sick contacts, immunised, no travel, normal growth

Case 1


Intrathoracic Mass
Anterior Mediastinum
Middle Mediastinum
Posterior Mediastinum
Lung
Chest Wall / Pleura
Lymphoma
Germ Cell Tumour (Teratoma)
Thymic
Lymphadenopathy
Foregut Cyst / Malformation
Vascular Lesion
Neurogenic Tumour
Neuroblastoma / Ganglioneuroma
Neurofibroma
Congenital
Tumour
Infection
Inflammatory mass
Sarcoma
Benign Neoplasm
Congenital anomaly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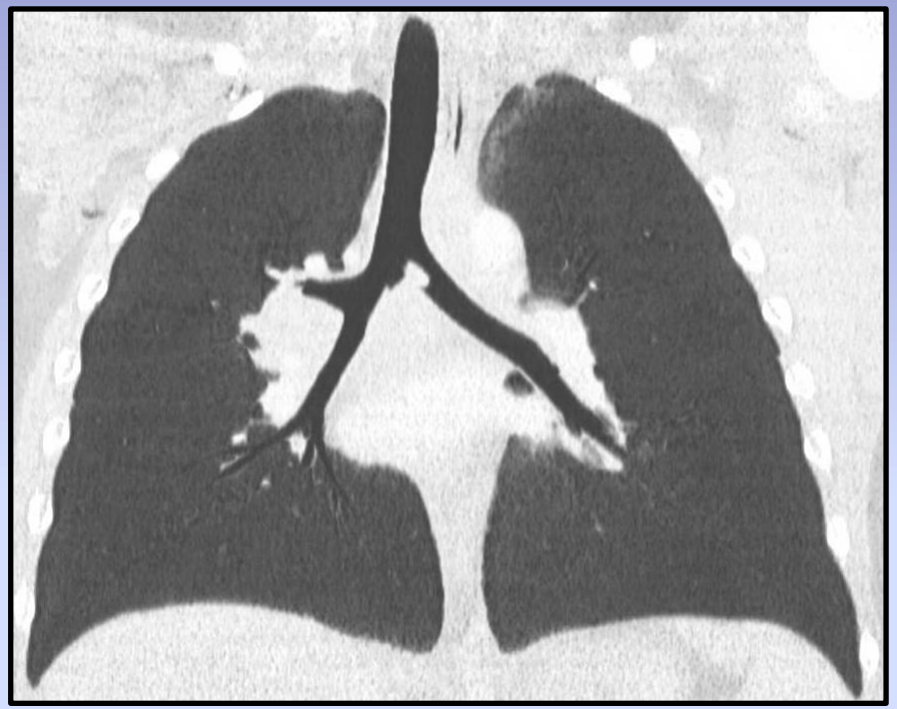
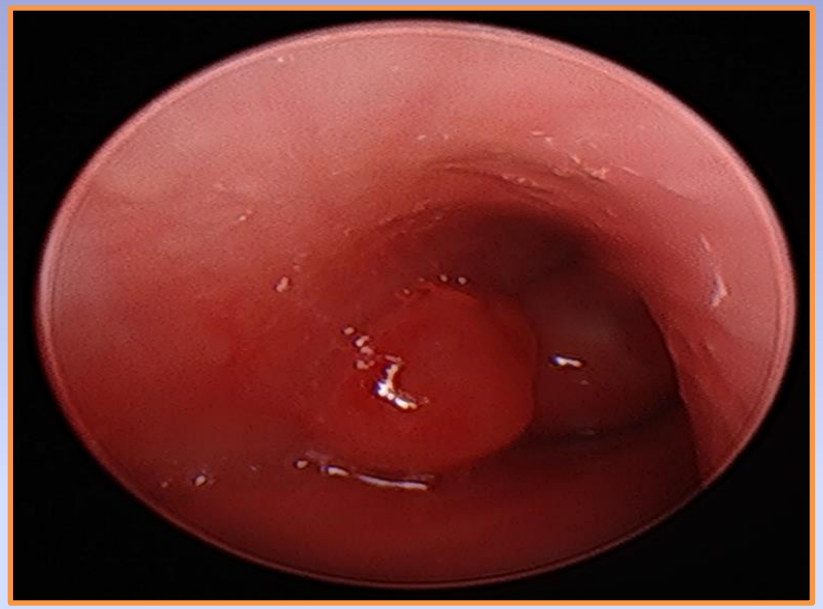
Case 1



- Malignancy: (e.g. carcinoma), metastatic
- Benign: hamartoma, myofibroma, papilloma
- Carcinoid
- Vascular: Haemangioma
- Inflammatory: granulomatous lesions
- Infective: bacterial, mycobacterial (e.g. tuberculosis), fungal (e.g. histoplasmosis, cryptococcus)

Case 1



- Bronchoscopy with ENT and resp teams
- Biopsy of nodule sent along with extensive bloodwork

- Vasculitic screen, immunodeficiency workup, all infective serology/PCR, bronchial fluid NAD
- Biopsy showed necrotizing granuloma, no neoplastic change
- Pt recovered well; no wheeze
- 5 weeks later: growth of mycobacterium avium complex from tissue biopsy

Case 1

Diagnosis: bronchial / mediastinal MAC

- Thoracic MAC infection is generally opportunistic, occurring in heavily immunocompromised patients
- Mendelien susceptibility to mycobacterial disease is a rare set of genetic conditions that can present in young children with invasive mycobacterial disease only (including from BCG vaccine)
- Intrathoracic (mediastinal & endobronchial) MAC infections in immunocompetent children have been reported in multiple case series and case reports; treatment consists of multi-drug therapy +/- surgical resection before

Endobronchial	2/M	*Mycobacterium avium/ M. intracellulare* complex	Partial resection; INH, Rif (14 mo)	Cured
Endobronchial	1/F	*M. avium/M. intracellulare* complex	Partial resection; INH, Rif (16 w)	Cured
Endobronchial, mediastinal	0.5/M	*M. avium/M. intracellulare* complex	Partial surgical resection	Cured
Endobronchial	2/M	*M. avium/M. intracellulare*	Surgical resection; INH, Rif, PZA (4 mo)	Cured
Mediastinal	1/F	*M. avium/M. intracellulare*	INH, Rif, PZA (6 mo)	Cured
Mediastinal	1/M	*M. avium* complex	INH, Rif (2 mo); surgical resection	Cured

- We treated our patient with ethambutol, rifampicin and azithromycin; he remained asymptomatic throughout

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Case 2

History

15 year old girl (55kg) presents to local emergency room:

- Fell and landed back 5 days prior; developed back pain
- Few days of high fever and sweats, mild headache, intermittent shortness of breath, tiredness and worsening feeling of general unwellness

Past / Family

- Anxiety and associated fatigue dx. 6mths prior; escitalopram
- Lives with two well younger siblings, parents, cat & dog
- No recent travel or sick contacts

Examination

- 38°C, tachycardic 130bpm, RR 18, saturations 98% RA
- Slightly delayed capillary refill
- Clear chest, normal heart sounds, soft abdomen

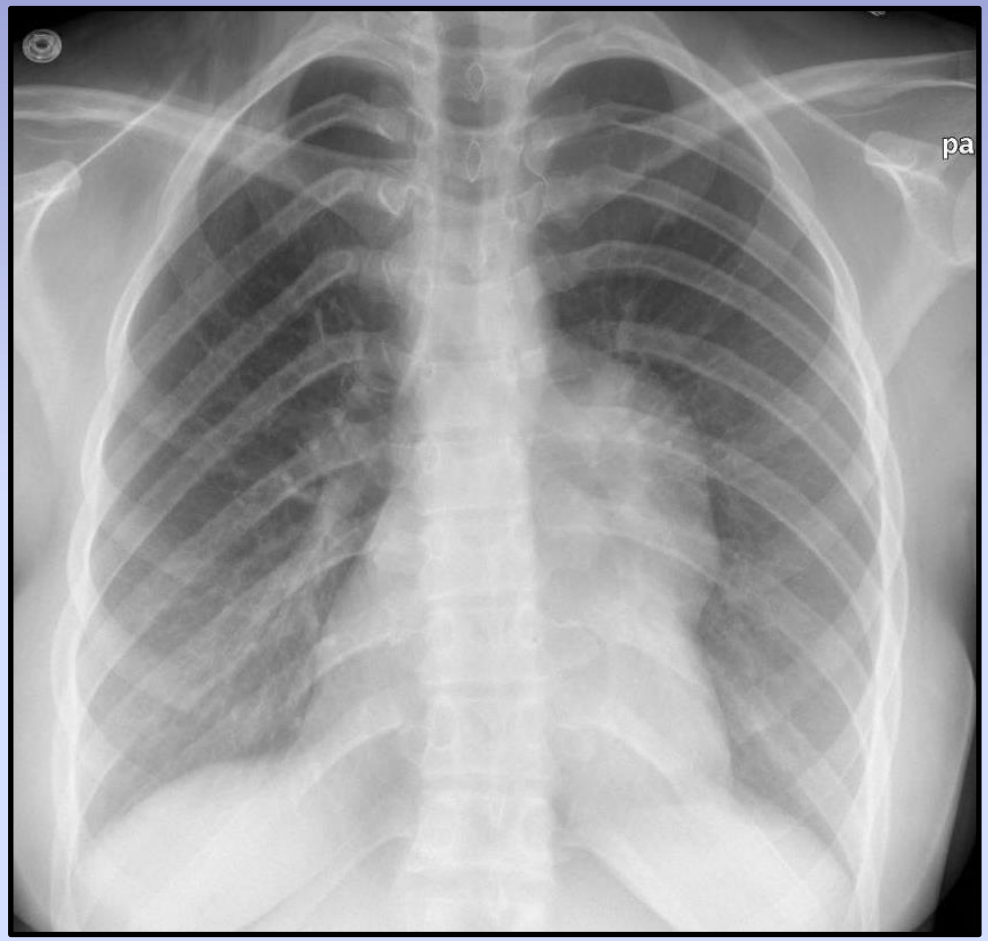
Case 2

Initial Investigations

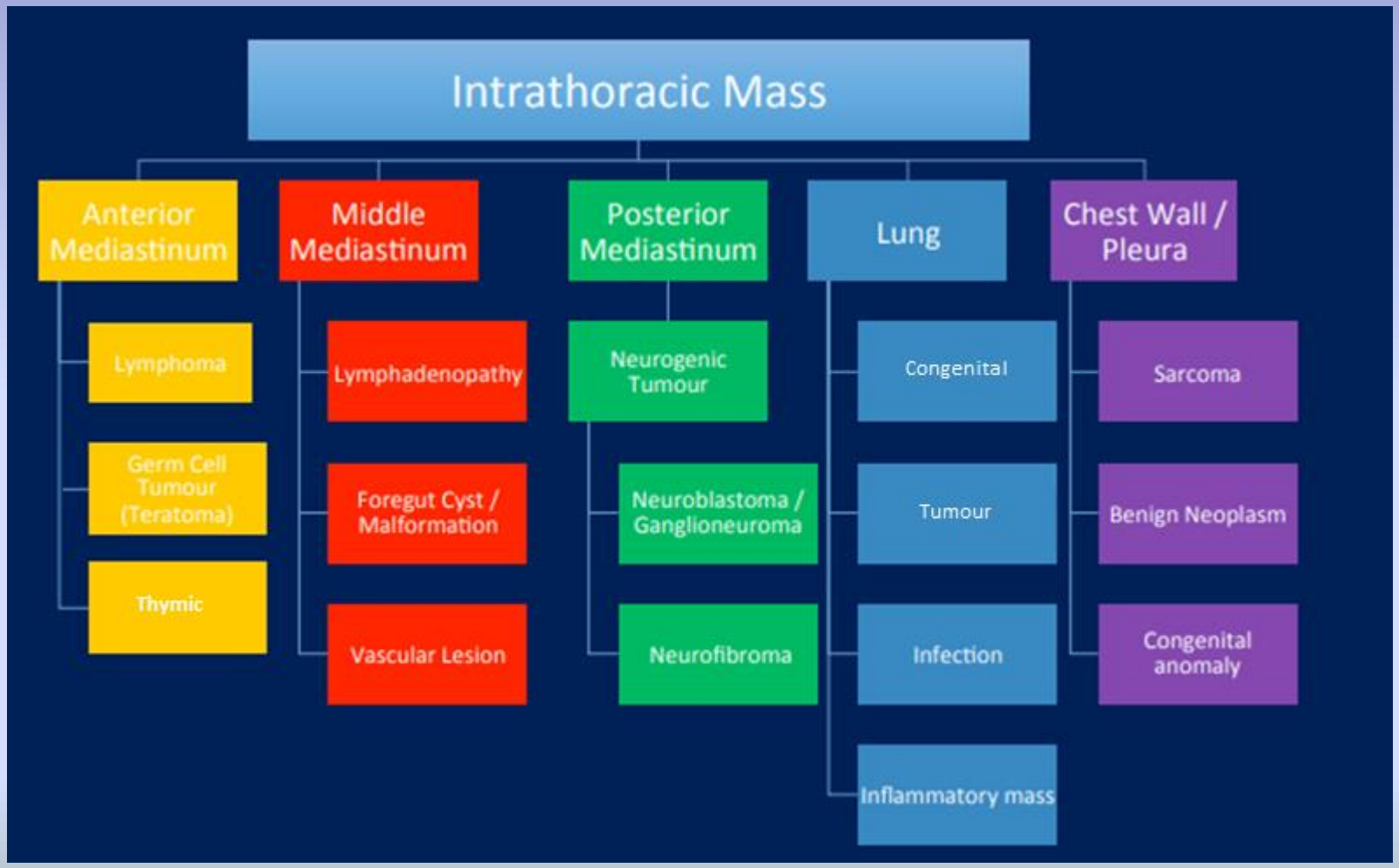
- WCC 19 (neutrophilia)
- CRP 365
- Creatinine 120
- Urinalysis: high WCC

Initial Management

- Treated for urosepsis and AKI; IV abx, fluid resuscitation: <2.5L
- Developed worsening shortness of breath



Case 2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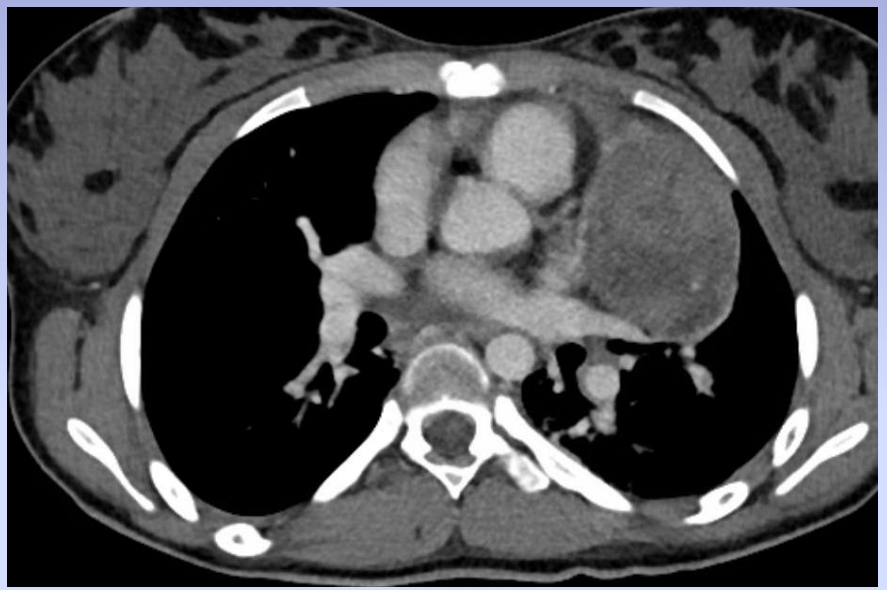
Case 2



Consultations

- Neurology
- Respiratory

Case 2

Further history

- 6 months of progressively worsening fatigue that seemed to be better in the morning but worse as the day progresses
- Increasing shortness of breath on exertion
- Couple of episodes of choking on food / difficulty with swallowing saliva
- Sleeping for long hours
- Dropped out of school, doing classes online
- Gave up soccer, finds walking short distances difficult
- Developed left droopy eyelid when tired
- A couple of episodes of slurred speech
- S/B ENT & psychologist who felt this was anxiety-related / somatic in origin

Further examination

- RR18, sats 97% RA, afebrile, HR 110bpm; clear chest, normal cardiac
- Symmetrical face and movements but bilateral eyelid ptosis
- Marked proximal muscle weakness, mildly unsteady gait
- Neck flexor muscle weakness
- Normal pupils, eye movements, tongue movements, gag
- Sensation and cognition appear intact
- Ice is applied to her left eyelid and there is marked improvement in ptosis

?

Case 2

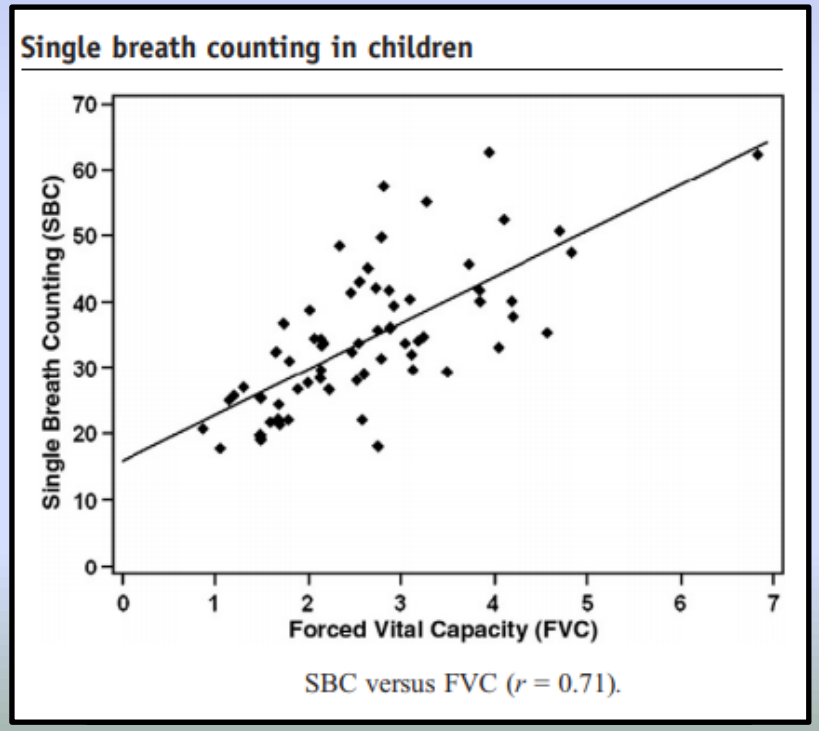
Provisional diagnosis: Myasthenia gravis secondary to antibody-producing thymoma

Investigations:

- Anti-MuSK and AchR antibodies sent
- Capillary gas: normal
- **SBCT score of 7**
- **Bedside slow VC: 480ml**

<u>Recommendations</u>

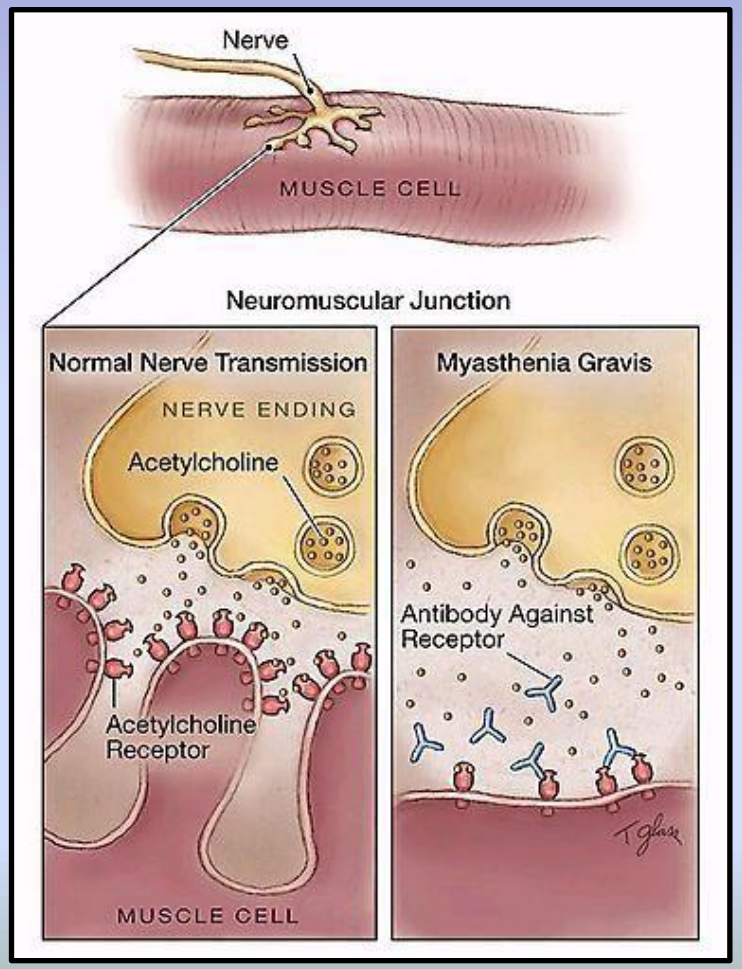
- **ICU review for consideration of respiratory support**
- **Immediate treatment with an acetylcholinesterase inhibitor (in liaison with neurology team)**



SBC versus FVC ($r = 0.71$).

Case 2

Pathophysiology



Assessment of severity

Most useful

- Trends of slow VC, MIP and (to a lesser extent) SBCT
- Weak neck flexion (correlates with diaphragm dysfunction)
- Severely slurred speech, drooling / choking on secretions

Somewhat useful

- Single point cutoffs:
  - SBCT <10-20; significant resp. muscle weakness
  - MIP <20-30cm H2O or VC <10-20ml/kg strongly suggests need for NIPPV / I+V

Less useful

- Use of accessory muscles
- Blood gases and oximetry

Case 2

Triggers for myasthenic crisis / deterioration

- Surgery
- Infection
- Tapering of immunosuppression
- Sleep deprivation
- Heat
- Pain
- Emotional distress
- Pregnancy / childbirth
- Drugs (see table)

Table 1 Drugs Producing Worsening of Myasthenic Weakness

Neuromuscular blocking agents
Antibiotics
Aminoglycosides, particularly gentamycin
Macrolides, particularly erythromycin and azithromycin
Cardiovascular agents
Beta-blockers
Calcium channel blockers
Procainamide
Quinidine
Quinine
Corticosteroids
Magnesium salts
Antacids, laxatives, intravenous tocolytics
Iodinated contrast agents
D-penicillamine

Case 2

Progress

- Rapid improvement with initial dose of pyridostigmine (Slow VC 0.48 → 1.3L and steadily improved to normal)
- Initial treatment involved continuing pyridostigmine, oral prednisone and plasmapheresis
- Antibody testing showed AchR antibodies
- EMG showed characteristic MG pattern also
- Biopsy confirmed the diagnosis of a type B1 (lymphocytic) thymic tumour (good prognosis)
- Pt. underwent total surgical resection 10 days ago, cx by post-op haemothorax requiring chest drain insertion; subsequent resolution & d/c from hospital 6 days ago
- Continues on pyridostigmine; awaiting neurology review

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Questions / Comments

Quokka

Bilby

Blue ringed octopus

Tasmanian Devil

Red back spider

Black flying fox