required repeat procedures. Underlying condition was diverse (21 conditions): Secundum ASD 19/88 (21.6%), Fallot's Tetralogy 19/88 (21.6%), and Ebstein's anomaly 9/88 (10.2%) were the most common.

Conclusions Whilst arrhythmias are most common in Secundum ASD, complex ACHD (ccTGA, complex congenital, TGA and Ebstein's anomaly) have a disproportionately high arrhythmia burden. 10% increase in EP procedures noted compared to our 2006 study. This is appropriate for to avoid recurrent admissions and long term anti-arrhythmics. Complex anatomy, challenging venous access and scar related re-entry tachycardias underline the need to expand the subspecialist EP-CHD capacity internationally.

18 KAWASAKI DISEASE- UNUSUAL FULMINANT PRESENTATION IN A SIX WEEK OLD INFANT WITH SYSTEMIC AND CARDIAC INVOLVEMENT, DIAGNOSIS AND MANAGEMENT CONUNDRUMS

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A six week infant was admitted with three days history of coryza and high grade temperature. She received treatment as presumed sepsis after sending full septic screen, congenital viral and retroviral serologies. She had high inflammatory markers along with raised liver enzymes and thrombocytopenia. She did not return any positive microbiology as well as viral serologies apart from persistently high inflammatory markers.

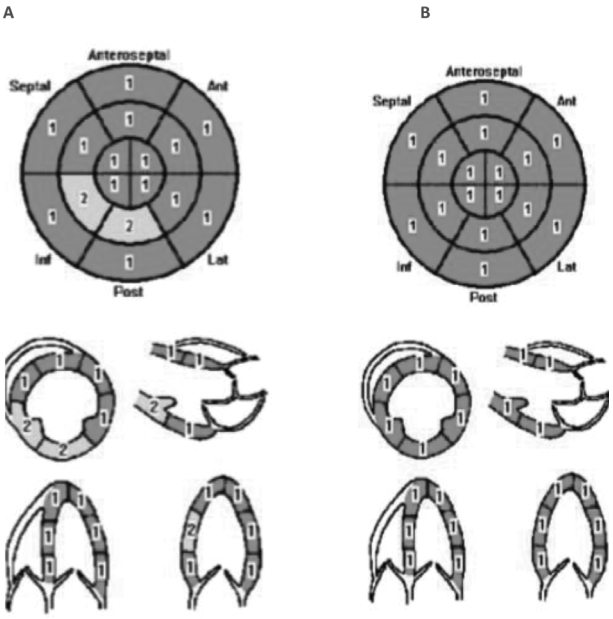
On day five of illness, she developed vasculitic changes in eyes, mouth and extremities concurrently with infiltrative pulmonary disease which required non invasive ventilatory support, hepatitis, renal dysfunction, anaemia, hypoalbuminaemia, hypofibrinogenaemia, raised ferritin, raised triglycerides in addition to thrombocytopenia. Initial echocardiogram was normal.

She was transferred to a tertiary paediatric centre for further support with rheumatology and immunology teams. She had been started on treatment for Kawasaki disease with aspirin and immunoglobulins. A repeat echocardiogram at day seven of illness showed evidence of coronary artery dilatation.

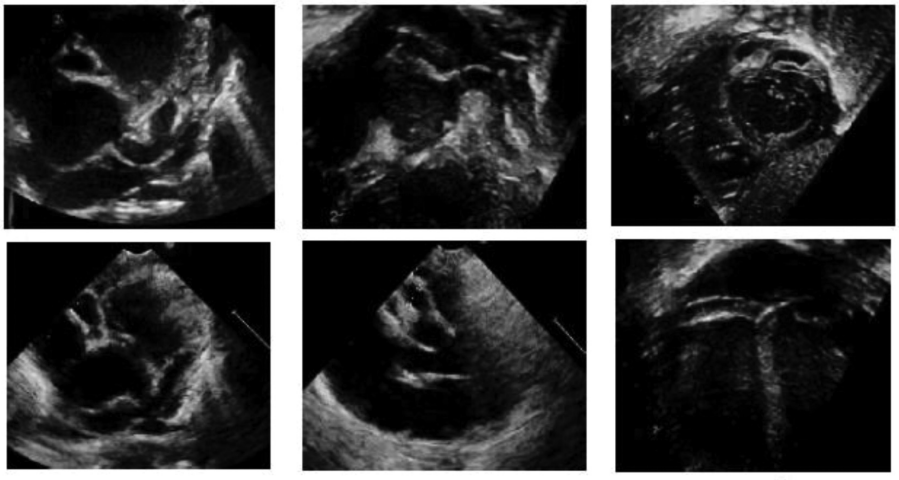
In view of ongoing severe inflammatory changes at two weeks of illness, she received a further course of immunoglobulins along with a course of methylprednisolone and then infliximab afterwards.The coronary artery dilatation had worsened with Z scores of 9–10 in all three coronaries. The skin biopsy was done because of profound vasculitic changes and peeling skin, although showed abnormalities, was not conclusive.

She was then transferred to the intensive care unit in our regional cardiac centre where she was treated with therapeutic heparin infusion then subcutaneous heparin and aspirin and further stabilised before she was successfully discharged home.

Kawasaki treatment guidelines in refractory disease has been updated in our unit.



Abstract 18 Figure 2 A&B Wall motion abnormality scoring 7 weeks (A) and 15 weeks (B) after peek presentation (A).



Abstract 18 Figure 1 Echocardiogram images of Coronary Arteries @ at peak of presentation