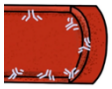
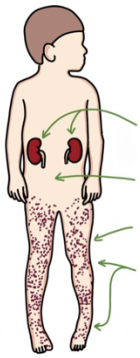
PAEDIATRIC RHEUMATOLOGY

	Rheumatic Fever	Juvenile Idiopathic Arthritis (JIA)	Ehler-Danlos Syndrome (EDS)	HSP (IgA vasculitis)	Kawasaki
PP	**Type 2 hypersensitive autoimmune** condition triggered by antibodies against GAS ➢ 2-4 weeks after initial infection (e.g. tonsillitis, pharyngitis) ***Multi-system disorder affecting*** ➢ Heart ➢ Joints, skin	**Autoimmune inflammation in joints** (i.e. arthritis without any cause) ***Types*** ➢ **Systemic JIA (Still's disease)** ➢ Polyarticular JIA ➢ Oligoarticular JIA ➢ Enthesitis -related arthritis ➢ Juvenile psoriatic arthritis	Umbrella term for **genetic collagen defects causing hypermobility in CT** ➢ Autosomal dominant ***Types*** ➢ Hypermobile EDS ➢ Classical EDS ➢ Vascular EDS ➢ Kyphoscoliotic EDS	**IgA deposits** in blood vessels affecting: ➢ **Skin** ➢ **Kidneys** ➢ **GIT** ***Must exclude*** ➢ Meningococcal ➢ Sepsis ➢ Acute Leukemia ➢ HUS ➢ ITP	• Mucocutaneous LN syndrome • systemic medium sized vessel vasculitis
RF	• ATSI • Female • Low SES or overcrowded • Ages 5-14	FHx of autoimmune DDx: *septic, osteomyelitis, osteosarcoma, transient, Perthes*			• Children < 5yo • Boys
Sx	***JONES criteria*** ***(2 major OR 1 major + 2 minor)*** ***Major*** • **Joints** – Migratory painful inflammatory polyarthritis • **Organ inflamed** (endocarditis - ↑HR, pericardial rub, HF) • **N** - SC nodules (firm painless nodulues) • **E** – erythema marginatum (pink rings on torso and proximal limb) • **S** – Sydenham chorea (XS dopamine releasee – invol. Jerky movements) **Minor** • Fever • ECG changes (prolonged PR) • Arthralgia w/o arthritis • Raised CRP/ESR	**More than > 6 weeks Sx in patients under age of 16** ➢ Joint pain ➢ Swelling ➢ Stiffness ***Systemic JIA*** ➢ Salmon-pink rash ➢ Swinging fevers (> 5day) ➢ Joint pain ➢ Enlarged LN ➢ UWL ***Polyarticular JIA (> 5 joints)*** ➢ Idiopathic symmetrical ➢ Mild fever, anaemia, and reduced growth ***Oligoarticular JIA (≤ 4 large joints)*** ➢ Anterior uveitis ➢ ANA +ve ***Enthesitis-Related Arthritis*** ➢ Seronegative – AS, RA, IBD, psoriatic ➢ Enthesitis → IP joints, wrist, ASIS, Achilles, patellar insertion ***Juvenile Psoriatic Arthritis*** ➢ Symmetrical polyarthritis affecting small joints OR asymmetrical large joints ➢ *Psoriasis, pitting, onycholysis, dactylitis, psoriasis plaques*	*Hypermobile EDS* ➢ Hypermobility of joints ➢ Soft stretchy skin *Classical EDS* ➢ Velvet smooth skin ➢ Abnormal wound healing ➢ High risk of prolapse, hernias, MR and aortic root dilatation *Vascular EDS* ➢ Thin, translucent skin ➢ Fragile BVs – spontaneous bleed *Kyphoscoliotic EDS* ➢ Hypotonia when neonate ➢ Kyphoscoliosis when growing older ➢ Rupture of medium sized arteries (GIT, coronary)	• **Non-blanching purpura rash** on buttocks and lower limbs (100%) - inflamed and leaky small BVs • **Abdo pain** (50%) • **Arthralgia** (75%) • **Haematuria** - IgA vasculitis (50%) 	• Persistent high fever > 5 days ***CREAM*** • *Bilateral* **Conjunctivitis** • Widespread red Blanching maculo-papular **rash** • **Oedema** • **Adenopathy** • **Muco-cutaneous** - cracked lips and strawberry tongue -*red tongue w/ large papillae* skin desquamation *Disease course* ➢ **Acute phase** – unwell + rash, fever, LN (for 1-2 wks) ➢ **Subacute phase** – arthralgia, skin desquamation (2-4 wks) ➢ **Convalescent (recovery) period** – bloods return normal and aneurysms may regress (2-4 weeks)
Comp.	• Recurrent rheumatic fever • Structural Heart – **mitral stenosis (>10yo) and IE**, • Arrythmias – AF • Chronic HF • PSGN (weeks after)	Macrophage activation system (MAS) – severe immune response presenting as ➢ DIC ➢ Low Hb, Low PLTS ➢ Non-blanching rash ➢ **Low ESR**	***Early onset OA*** ***POTS*** – due to ANS dysfunction ➢ Inappropriate tachycardia on sitting or standing up ➢ Pre-sycope, headache, nausea	• **ISS** • Bowel infraction • Gi haemorrhage • IgA nephritis	Coronary artery aneurysm
Ix	• Throat swab • ASOT +ve • ECHO – carditis, IE • ECG – AF? • CXR • UA	**Clinical Dx** ➢ Raised CRP, ESR ➢ Raised serum ferritin ➢ ANA ➢ RF, anti-CCP	**Beighton score (/9) for hypermobility to Dx** ➢ Elbows/knee hypertEXT ➢ Thumb bend to forearm ➢ Pinky extends > 90 degs **DDx: Marfan's – higharched palette, archnodactyly, arm span**	• BP - ?HTN • FBC, +blood film - ?sepsis, leukemia • EUC → ?nephrotic • CRP → ?sepsis • Blood cultures • Urine dipstick + ACR	• FBC – anemia, ↑ WCC, ↑ plt, • LFT – low albumin, ↑ LFT • ESR - raised • Urinalysis – pyuria with no infection • ECHO
Mx	***Main Rx:*** Penicillin V (phenoxy-methylpenicillin) for 10 days ***Supportive Rx:*** • Hand hygiene • Analgesia (NSAID) – athralgia • Aspirin + steroids – for carditis • Prophylactic ABx – PO or IM (continued into adulthood) • Valproate – Sydenham chorea • CHADS-VASC ≥ 1 = DOAC	***Referral*** • Paed rheumatology • MDT involvement • Ophthal referral if oligoarticular JIA ***Supportive Rx*** to reduce joint inflammation and minimise Sx: ➢ **NSAID** (ibuprofen) ➢ **IM/IA steroids** (oligoarthritis) ➢ **DMARDs** (MTX, SSZ, leflunomide) ➢ **Biologic** (TNFa inhibitor – infliximab, adalimumab, etanercpt)	**NO CURE!!** **Aim to maintain healthy joints and stop complications** ➢ F/U – rheum ➢ PT -stabilise joints ➢ OT -max function + maintain good joint posture ➢ Psychology - QoL	***Supportive Mx*** • Analgesia • Rest • Hydration • Abdo resolves within few days ***Follow up on:*** • Urine dipstick for proteinuria and haematuria • Regular BP – HTN • Warn recurrence in 6/12 for 33% of patients	*Urgent paeds referral* ➢ **High dose aspirin** to reduce risk of thrombosis ➢ **IVIg** to reduce risk of coronary artery aneurysms ➢ F/U ECHO after recovery **Beware of Reye's syndrome** = LE swelling of liver and brain → confusion, seizures, LOC *__ONLY__ condition where aspirin is used in children

Nb: Fevers for more than 5 days, the key non-infective differentials to remember are Kawasaki disease, Still's disease, rheumatic fever and leukaemia.