Centre……..……..Assessor…….Date……

Patient name…….Patient code……*PVAS 2008 Study*

PAEDIATRIC VASCULITIS ACTIVITY SCORE 2008

○ Tick "**Active**" box **only** if abnormality due to active vasculitis is newly present or worse over the last 4 weeks or persists for less than 3 months. After that, if ALL items are persistent and represent smouldering/low grade/grumbling disease, and there are no new/worse features, please tick the box at the bottom right corner. At the very first assessment all active items are considered as active/worse. If there are no abnormalities in a system, please tick the "**None**" box. For items present longer than 3 months refer to the Vasculitis Damage Index to score damage.

	None	Active
1. General	○	
Myalgia		○
Arthralgia or arthritis		○
Fever ≥ 38.0 ^{0}C		○
Weight Loss ≥ 5% body weight		○
2. Cutaneous	○	
Polymorphous exanthema		○
Livedo		○
Panniculitis		○
Purpura		○
Skin nodules		○
Infarct (nail edge lesion, splinter haemorrhage)		○
Ulcer (full-thickness necrosis)		○
Gangrene (extensive necrosis)		○
Other skin vasculitis (specify below)		○
3. Mucous membranes/eyes	○	
Mouth ulcers/granulomata		○
Genital ulcers		○
Adnexal inflammation		○
Significant proptosis		○
Red eye (Epi)scleritis		○
Red eye conjunctivitis/ blepharitis/keratitis		○
Uveitis		○
Blurred vision		○
Sudden visual loss		○
Retinal vasculitis/retinal vessel thrombosis/ retinal exudates/haemorrhages		○
4. ENT	○	
Nasal discharge/crusts/ulcers/granuloma		○
Paranasal sinus involvement		○
Subglottic stenosis/ hoarseness /stridor		○
Conductive hearing loss		○
Sensorineural hearing loss		○
5. Chest	○	
Wheeze or expiratory dyspnea		○
Endobronchial/endotracheal involvement		○
Nodules or cavities		○
Pleural effusion/pleurisy		○
Infiltrate		○
Massive haemoptysis/Alveolar haemorrhage		○
Respiratory failure		○

	None	Active
6. Cardiovascular	○	
Loss of pulses		○
Bruits over accessible arteries		○
Blood pressure discrepancy		○
Claudication of extremities		○
Ischaemic cardiac pain		○
Cardiomyopathy		○
Congestive cardiac failure		○
Valvular heart disease		○
Pericarditis		○
7. Abdominal	○	
Abdominal pain		○
Peritonitis		○
Blood in stools or bloody diarrhoea		○
Bowel ischaemia		○
8. Renal	○	
Hypertension >95th centile (for height)		○
Proteinuria >0.3 g/24h,>20mmol/mg creatinin		○
Haematuria ≥2+ or 5 rbc/hpf or red cell casts		○
GFR 50-80ml/min/1.73 m^2		○
GFR 15-49 ml/min/1.73 m^2		○
GFR <15 ml/min/1.73m^2		○
Rise in creatinine > 10% or		
Creatinine clearance (GFR) fall > 25%		○
9. Nervous system	○	
Headache		○
Meningitis/encephalitis		○
Organic confusion/cognitive dysfunction		○
Seizures (not hypertensive)		○
Stroke		○
Cord lesion		○
Cranial nerve palsy		○
Sensory peripheral neuropathy		○
Motor mononeuritis multiplex		○
10. OTHER	○	
		○
		○
NO NEW/WORSE DISEASE :		
Tick here if there is no new/worse abnormality present in ANY of the systems above and active items represent low grade grumbling disease		

Modified from Luqmani R.A., Bacon P.A., Moots R.J. et al (1994) BVAS in systemic necrotizing vasculitis. Quart J Med 87: 671-8

Glossary and scoring for PVAS. GENERAL RULE: disease features are scored only when they are due to active vasculitis, after excluding other causes (e.g. infection, hypertension, etc.). If the feature is due to active disease, it is scored in the boxes. It is essential to apply these principles to each item below. Scores have been weighted according to the severity which each symptom or sign is thought to represent. Tick "Persistent Disease" box if all the abnormalities are due to active (but not new or worse) vasculitis. If any of the abnormalities are due to new/worse disease , DO NOT tick the "Persistent Disease" box. For some features, further information (from specialist opinion or further tests) is required if abnormality is newly present or worse. Remember that in most instances, you will be able to complete the whole record when you see the patient. However, you may need further information before entering some items. Please leave these items blank, until the information is available, and then fill them in. For example, if the patient has new onset of stridor, you would usually ask an ENT colleague to investigate this further to determine whether or not it is due to active Wegener's granulomatosis.		**PVAS persistent**	**PVAS new/ worse**
1. General	**Maximum scores**	**2**	**3**
Myalgia	Diffuse, spontaneous, hard to localize muscle pain or tenderness on muscle palpation. Exclude fibromyalgia.	1	1
Arthralgia or arthritis	Joint pain in any number of joints or presence of objective signs of active synovitis: intraarticular swelling due to synovial proliferation and/or joint effusion with limited range of movement and/or pain on movement or joint tenderness. Any number of joints.	1	1
Fever ≥ 38.0 °C	Documented temperature elevation >38°C. The value refers to axillary/oral temperature (rectal temperature 0.5 °C higher). Exclude infections by appropriate cultures, serology and PCR methods.	2	2
Weight Loss ≥ 5% body weight	At least 5% loss of body weight (not fluid) having occurred since last assessment or in the 4 weeks not as a consequence of dieting	2	2
2. Cutaneous	**Maximum scores**	**3**	**6**
Polymorphous exanthema	Non-haemorrhagic, non-necrotising skin eruption of any type or combined types. Exclude allergy/drug reaction/infection	1	1
Livedo	Purplish reticular pattern usually irregularly distributed around subcutaneous fat lobules, often more prominent with cooling, common over foot margins. Exclude antiphospholipid syndrome.	1	1
Panniculitis	Single or multiple tender deep subcutaneous nodules caused by inflammation of deep subcutaneous tissue with typical histopathology findings if biopsy performed	1	1
Purpura	Petechiae (small red spots), palpable purpura, or ecchymoses (large plaques) in skin or oozing (in the absence of trauma) in the mucous membranes.	1	2
Skin nodules	Subcutaneous nodules, often along arteries, tender on palpation.	1	1
Infarct	Nail edge lesion, splinter haemorrhage or flea bite lesion of small vessel vasculitis	1	1
Ulcer	Area of full-thickness skin/subcutaneous tissue ulceration/necrosis	1	4
Gangrene	Extensive skin/subcutaneous tissue/underlying structure necrosis, digital phalanx or other peripheral (nose, ear tips) necrosis/gangrene	2	6
Other skin vasculitis	Vasculitis different from previous e.g. subcutaneous swelling/oedema due to capillary leak in small vessel involvement, Raynaud's phenomenon etc.	1	1
3. Mucous membranes/eyes	**Maximum scores**	**3**	**6**
Mouth ulcers/granulomata	Aphtous stomatitis, ischaemic ulcers and/or granulomatous inflammation in oral cavity. Exclude other causes (SLE, infection)	1	2
Genital ulcers	Ulcers localised in the genitalia or perineum, excluding infections.	1	1
Adnexal inflammation	Salivary (diffuse, tender swelling unrelated to meals) or lacrimal gland inflammation. Exclude other causes (infection). Specialist opinion preferably required.	2	4
Significant proptosis	Protrusion of the eyeball due to significant amounts of inflammatory in the orbit; if unilateral, there should be a difference of 2 mm between one eye and the other. This may be associated with diplopia due to infiltration of extra-ocular muscles. Developing myopia (measured on best visual acuity, see later) can also be a manifestation of proptosis	2	4
Red eye (Epi)scleritis	Inflammation of the sclerae (specialist opinion usually required). Can be heralded by photophobia.	1	2
Red eye conjunctivitis Blepharitis Keratitis	Inflammation of the conjuctivae (exclude infectious causes and excluding uveitis as cause of red eye, also exclude conjunctivitis sicca which should not be scored as this is not a feature of active vasculitis); (specialist opinion not usually required). Inflammation of eyelids. Exclude other causes (trauma, infection). Usually no specialist opinion is required Inflammation of central or peripheral cornea as evaluated by specialist	1	1
Blurred vision	Altered measurement of best visual acuity from previous or baseline, requiring specialist opinion for further evaluation.	2	3
Sudden visual loss	Sudden loss of vision requiring ophthalmological assessment.		6
Uveitis	Inflammation of the uvea (iris, ciliary body, choroid) confirmed by ophthalmologist.	2	6
Retinal vasculitis	Retinal vessel sheathing on examination by specialist or confirmed by retinal fluoroscein angiography	2	6
Retinal vessel thrombosis	Arterial or venous retinal blood vessel occlusion		
Retinal exudates	Any area of soft retinal exudates (exclude hard exudates) seen on ophthalmoscopic examination.		
Retinal haemorrhages	Any area of retinal haemorrhage seen on ophthalmoscopic examination.		
4. ENT	**Maximum scores**	**3**	**6**
Bloody nasal discharge/ nasal crusts/ulcers and/or granulomata	Bloody, mucopurulent, nasal secretion, light or dark brown crusts frequently obstructing the nose, nasal ulcers and/or granulomatous lesions observed by rhinoscopy	2	4
Paranasal sinus involvement	Tenderness or pain over paranasal sinuses usually with pathologic imaging (CT, MR, x-ray, ultrasound)	1	2
Subglottic stenosis	Stridor and hoarseness due to inflammation and narrowing of the subglottic area observed by laryngoscopy	3	6
		PVAS persistent	**PVAS new/worse**
Conductive hearing loss	Hearing loss due to middle ear involvement confirmed by otoscopy and/or tuning fork examination and/or audiometry	1	3

Original published in Arthritis Research & Therapy 2015, 17:292 doi:10.1186/s13075-015-0810-3.