

# PAEDIATRIC VASCULITIS ACTIVITY SCORE *Example 1*

○✗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 Paediatric Vasculitis Damage Index to score damage.

	None	Active		None	Active
<b>1. General</b>	○		<b>6. Cardiovascular</b>	✗	
Myalgia		○	Loss of pulses		○
Arthralgia or arthritis		✗	Bruits over accessible arteries		○
Fever ≥ 38.0 °C		○	Blood pressure discrepancy		○
Weight Loss ≥ 5% body weight		✗	Caudication of extremities		○
			Ischaemic cardiac pain		○
<b>2. Cutaneous</b>		✗	Cardiomyopathy		○
Polymorphous exanthema		○	Congestive cardiac failure		○
Livedo		○	Valvular heart disease		○
Panniculitis		○	Pericarditis		○
Purpura		○	<b>7. Abdominal</b>		✗
Skin nodules		○	Abdominal pain		○
Infarct (nail edge lesion, splinter haemorrhage)		○	Peritonitis		○
Ulcer (full-thickness necrosis)		○	Blood in stools or bloody diarrhoea		○
Gangrene (extensive necrosis)		○	Bowel ischaemia		○
Other skin vasculitis (specify below)		○			
<b>3. Mucous membranes/eyes</b>		✗	<b>8. Renal</b>		○
Mouth ulcers/granulomata		○	Hypertension >95th centile (for height)		○
Genital ulcers		○	Proteinuria >0.3 g/24h,>20mg/mmol creatinine		○
Adnexal inflammation		○	Haematuria ≥2+ or 5 rbc/hpf or red cell casts		✗
Significant proptosis		○	GFR 50-80ml/min/1.73 m <sup>2</sup>		○
Red eye (Epi)scleritis		○	GFR 15-49 ml/min/1.73 m <sup>2</sup>		○
Red eye conjunctivitis/ blepharitis/keratitis		○	GFR <15 ml/min/1.73m <sup>2</sup>		○
Uveitis		○	Rise in creatinine > 10% or		
Blurred vision		○	Creatinine clearance (GFR) fall > 25%		✗
Sudden visual loss		○	<b>9. Nervous system</b>		✗
Retinal vasculitis/retinal vessel thrombosis/ retinal exudates/haemorrhages		○	Headache		○
			Meningitis/encephalitis		○
<b>4. ENT</b>		○	Organic confusion/cognitive dysfunction		○
Nasal discharge/crusts/ulcers/granuloma		✗	Seizures (not hypertensive)		○
Paranasal sinus involvement		✗	Stroke		○
Subglottic stenosis/ hoarseness /stridor		○	Cord lesion		○
Conductive hearing loss		○	Cranial nerve palsy		○
Sensorineural hearing loss		○	Sensory peripheral neuropathy		○
			Motor mononeuritis multiplex		○
<b>5. Chest</b>		✗	<b>10. OTHER</b>		○
Wheeze or expiratory dyspnea		○	Malaise		✗
Endobronchial/endotracheal involvement		○			○
Nodules or cavities		○			
Pleural effusion/pleurisy		○	<b>PERSISTENT DISEASE ONLY :</b>		
Infiltrate		○	Tick here if there is no new/worse abnormality present in ANY of the systems above and all active items represent persistent disease		<input type="checkbox"/>
Massive haemoptysis/Alveolar haemorrhage		○			
Respiratory failure		○			

# PAEDIATRIC VASCULITIS ACTIVITY SCORE *Example II*

○✗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 Paediatric Vasculitis Damage Index to score damage.

	None	Active		None	Active
<b>1. General</b>	○		<b>6. Cardiovascular</b>	✗	
Myalgia		○	Loss of pulses		○
Arthralgia or arthritis		○	Bruits over accessible arteries		○
Fever ≥ 38.0 °C		✗	Blood pressure discrepancy		○
Weight Loss ≥ 5% body weight		○	Caudication of extremities		○
			Ischaemic cardiac pain		○
<b>2. Cutaneous</b>		✗	Cardiomyopathy		○
Polymorphous exanthema		○	Congestive cardiac failure		○
Livedo		○	Valvular heart disease		○
Panniculitis		○	Pericarditis		○
Purpura		○	<b>7. Abdominal</b>		✗
Skin nodules		○	Abdominal pain		○
Infarct (nail edge lesion, splinter haemorrhage)		○	Peritonitis		○
Ulcer (full-thickness necrosis)		○	Blood in stools or bloody diarrhoea		○
Gangrene (extensive necrosis)		○	Bowel ischaemia		○
Other skin vasculitis (specify below)		○	<b>8. Renal</b>		○
<b>3. Mucous membranes/eyes</b>		✗	Hypertension >95th centile (for height)		✗
Mouth ulcers/granulomata		○	Proteinuria >0.3 g/24h,>20mg/mmol creatinine		✗
Genital ulcers		○	Haematuria ≥2+ or 5 rbc/hpf or red cell casts		✗
Adnexal inflammation		○	GFR 50-80ml/min/1.73 m <sup>2</sup>		○
Significant proptosis		○	GFR 15-49 ml/min/1.73 m <sup>2</sup>		○
Red eye (Epi)scleritis		○	GFR <15 ml/min/1.73m <sup>2</sup>		○
Red eye conjunctivitis/ blepharitis/keratitis		○	Rise in creatinine > 10% or		
Uveitis		○	Creatinine clearance (GFR) fall > 25%		○
Blurred vision		○	<b>9. Nervous system</b>		✗
Sudden visual loss		○	Headache		○
Retinal vasculitis/retinal vessel thrombosis/ retinal exudates/haemorrhages		○	Meningitis/encephalitis		○
<b>4. ENT</b>		○	Organic confusion/cognitive dysfunction		○
Nasal discharge/crusts/ulcers/granuloma		✗	Seizures (not hypertensive)		○
Paranasal sinus involvement		✗	Stroke		○
Subglottic stenosis/ hoarseness /stridor		○	Cord lesion		○
Conductive hearing loss		✗	Cranial nerve palsy		○
Sensorineural hearing loss		○	Sensory peripheral neuropathy		○
<b>5. Chest</b>		✗	Motor mononeuritis multiplex		○
Wheeze or expiratory dyspnea		○	<b>10. OTHER</b>		○
Endobronchial/endotracheal involvement		○	Malaise		✗
Nodules or cavities		○	Aural discharge		✗
Pleural effusion/pleurisy		○	<b>PERSISTENT DISEASE ONLY :</b>		
Infiltrate		○	Tick here if there is no new/worse abnormality present in ANY of the systems above and all active items represent persistent disease		<input type="checkbox"/>
Massive haemoptysis/Alveolar haemorrhage		○			
Respiratory failure		○			

# PAEDIATRIC VASCULITIS ACTIVITY SCORE *Example III*

○✗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 Paediatric Vasculitis Damage Index to score damage.

	None	Active		None	Active
<b>1. General</b>	✗		<b>6. Cardiovascular</b>	✗	
Myalgia		<input type="radio"/>	Loss of pulses		<input type="radio"/>
Arthralgia or arthritis		<input type="radio"/>	Bruits over accessible arteries		<input type="radio"/>
Fever $\geq 38.0^{\circ}\text{C}$		<input type="radio"/>	Blood pressure discrepancy		<input type="radio"/>
Weight Loss $\geq 5\%$ body weight		<input type="radio"/>	Caudication of extremities		<input type="radio"/>
			Ischaemic cardiac pain		<input type="radio"/>
<b>2. Cutaneous</b>	✗		Cardiomyopathy		<input type="radio"/>
Polymorphous exanthema		<input type="radio"/>	Congestive cardiac failure		<input type="radio"/>
Livedo		<input type="radio"/>	Valvular heart disease		<input type="radio"/>
Panniculitis		<input type="radio"/>	Pericarditis		<input type="radio"/>
Purpura		<input type="radio"/>	<b>7. Abdominal</b>	✗	
Skin nodules		<input type="radio"/>	Abdominal pain		<input type="radio"/>
Infarct (nail edge lesion, splinter haemorrhage)		<input type="radio"/>	Peritonitis		<input type="radio"/>
Ulcer (full-thickness necrosis)		<input type="radio"/>	Blood in stools or bloody diarrhoea		<input type="radio"/>
Gangrene (extensive necrosis)		<input type="radio"/>	Bowel ischaemia		<input type="radio"/>
Other skin vasculitis (specify below)		<input type="radio"/>			
<b>3. Mucous membranes/eyes</b>	✗		<b>8. Renal</b>	✗	
Mouth ulcers/granulomata		<input type="radio"/>	Hypertension $>95^{\text{th}}$ centile (for height)		<input type="radio"/>
Genital ulcers		<input type="radio"/>	Proteinuria $>0.3\text{ g}/24\text{h}, >20\text{mg}/\text{mmol}$ creatinine		<input type="radio"/>
Adnexal inflammation		<input type="radio"/>	Haematuria $\geq 2+$ or 5 rbc/hpf or red cell casts		<input type="radio"/>
Significant proptosis		<input type="radio"/>	GFR 50-80ml/min/ $1.73\text{ m}^2$		<input type="radio"/>
Red eye (Epi)scleritis		<input type="radio"/>	GFR 15-49 ml/min/ $1.73\text{ m}^2$		<input type="radio"/>
Red eye conjunctivitis/ blepharitis/keratitis		<input type="radio"/>	GFR $<15\text{ ml}/\text{min}/1.73\text{m}^2$		<input type="radio"/>
Uveitis		<input type="radio"/>	Rise in creatinine $> 10\%$ or		
Blurred vision		<input type="radio"/>	Creatinine clearance (GFR) fall $> 25\%$		<input type="radio"/>
Sudden visual loss		<input type="radio"/>	<b>9. Nervous system</b>	✗	
Retinal vasculitis/retinal vessel thrombosis/ retinal exudates/haemorrhages		<input type="radio"/>	Headache		<input type="radio"/>
			Meningitis/encephalitis		<input type="radio"/>
<b>4. ENT</b>	✗		Organic confusion/cognitive dysfunction		<input type="radio"/>
Nasal discharge/crusts/ulcers/granuloma		<input type="radio"/>	Seizures (not hypertensive)		<input type="radio"/>
Paranasal sinus involvement		<input type="radio"/>	Stroke		<input type="radio"/>
Subglottic stenosis/ hoarseness /stridor		<input type="radio"/>	Cord lesion		<input type="radio"/>
Conductive hearing loss		<input type="radio"/>	Cranial nerve palsy		<input type="radio"/>
Sensorineural hearing loss		<input type="radio"/>	Sensory peripheral neuropathy		<input type="radio"/>
			Motor mononeuritis multiplex		<input type="radio"/>
<b>5. Chest</b>	✗		<b>10. OTHER</b>	✗	
Wheeze or expiratory dyspnea		<input type="radio"/>			<input type="radio"/>
Endobronchial/endotracheal involvement		<input type="radio"/>			<input type="radio"/>
Nodules or cavities		<input type="radio"/>			
Pleural effusion/pleurisy		<input type="radio"/>	<b>PERSISTENT DISEASE ONLY :</b>		
Infiltrate		<input type="radio"/>	Tick here if there is no new/worse abnormality present in ANY of the systems above and all active items represent persistent disease		<input type="checkbox"/>
Massive haemoptysis/Alveolar haemorrhage		<input type="radio"/>			
Respiratory failure		<input type="radio"/>			

