

# Paediatric Vasculitis Damage Index Glossary

Disease damage is defined by **duration of the sign/symptom of at least 3 months** any time since the onset of vasculitis. Remember that the item does not need to be present at the time of scoring. Items currently present score under the column „**Present**“, items that had lasted for  $\geq 3$  months and have resolved by the time of evaluation score under the column „**NLP**“ (No Longer Present). Although some of the items and their description are very similar to those of disease activity score (PVAS), their DURATION and NEED FOR INCREASED IMMUNOSUPPRESSIVE TREATMENT are the main distinctions between the two.

<b>1. Musculoskeletal</b>	
<b>Significant muscle atrophy</b>	Demonstrated on clinical examination (not attributable to stroke because scored elsewhere), lasting $\geq 3$ months
<b>Deforming / erosive arthritis</b>	Persistent limitation of joint ROM or deformities on clinical examination, and/or erosions confirmed radiologically (excluding avascular necrosis), lasting $\geq 3$ months.
<b>Osteoporosis / vertebral collapse</b>	Vertebral collapse and/or insufficiency fractures confirmed with imaging to include bone densitometry (excluding avascular necrosis). Requires both evidence of decreased bone density and fracture; ‘low’ bone density by itself is insufficient.
<b>Avascular necrosis</b>	Confirmed by appropriate radiological imaging
<b>Chronic osteomyelitis</b>	Confirmed by radiological investigation and orthopaedic surgeon, lasting $\geq 3$ months.

<b>2. Skin / Mucous membranes</b>	
<b>Major alopecia</b>	Major (e.g. requiring wig) chronic hair loss with or without scars, documented clinically, lasting $\geq 3$ months.
<b>Chronic cutaneous ulcers, scars, striae</b>	Open sore on skin surface or significant scars from damage to dermis, including cutaneous striae, lasting $\geq 3$ months.
<b>Chronic mucosal ulcers</b>	Recurrent crops or persistent mouth/genital ulcers requiring therapy, lasting $\geq 3$ months. Microstomia resulting in restriction of mouth opening

<b>3. Ocular</b>	
<b>Cataract</b>	A lens opacity in one or both eyes documented by ophthalmoscopy.
<b>Chronic retinopathy</b>	Any significant retinal change on ophthalmoscopy (preferably confirmed and characterised by ophthalmologist); may result in field defect, or loss of acuity due to retinal vessel involvement present $\geq 3$ months; retinal scars
<b>Optic atrophy</b>	As seen on ophthalmoscopy in one or both eyes
<b>Chronic diplopia / dry eyes / photophobia</b>	Restricted eye movements (not due to nerve palsies), double vision or tunnel vision, loss of tears, likely from destructive dacrocystitis also contributing to photophobia
<b>Impaired visual acuity</b>	Vision impairment (better than 20/200 of Snellen chart or other relevant assessment methodology used) not correctable by refraction, assessed by a specialist

<b>Blindness (vision &lt; 20/200) in one eye</b>	Vision of 20/200 or less (Snellen chart or by other relevant assessment methodology used), not correctable by refraction and assessed by a specialist
<b>Blindness (vision &lt; 20/200) in second eye</b>	Vision of 20/200 or less (Snellen chart or by other relevant assessment methodology used), not correctable by refraction and assessed by a specialist
<b>Orbital wall destruction</b>	Significant bone destruction as documented on X-ray/CT/MRI.

<b>4. ENT</b>	
<b>Hearing loss</b>	Any hearing loss due to either middle ear involvement or to auditory nerve/ cochlear damage, confirmed by audiometry, lasting $\geq 3$ months.
<b>Chronic nasal blockage/ discharge/crusting</b>	Persistent difficulties breathing through the nose with purulent discharge and/or with crust formation usually requiring nasal lavage, lasting $\geq 3$ months
<b>Nasal bridge collapse/septal perforation</b>	Saddle nose deformity and/or perforation of nasal septum not related to other causes (cocaine or intranasal steroid use)
<b>Chronic sinusitis / bone destruction</b>	Chronic purulent nasal discharge and sinus pain with sinusitis lasting $\geq 3$ months or cavitating bone damage confirmed radiologically
<b>Chronic subglottic stenosis (no surgery)</b>	Persistent hoarseness and/or stridor confirmed by endoscopy and/or imaging lasting $\geq 3$ months, with or without local therapy excluding surgery
<b>Subglottic stenosis (with surgery)</b>	Persistent hoarseness and/or stridor confirmed by endoscopy and/or imaging, with surgery

<b>5. Chest</b> <i>NB lung function can only be reliably performed in children over 6 years of age.</i>	
<b>Chronic tracheal /endobronchial stenosis</b>	Confirmed by endoscopy, lasting $\geq 3$ months
<b>Chronic lower airway obstruction (PFT obstruction)</b>	Prolonged expiration with or without wheeze PLUS obstructive pattern on pulmonary function test (PFT), lasting $\geq 3$ months. Includes chronic asthma clinical picture, only if developing or worsening after vasculitis onset.
<b>Chronic breathlessness</b>	Tachypnea and/or breathing difficulties, often (but not necessarily) with restrictive pattern on PFT, lasting $\geq 3$ months, without confirmed pulmonary fibrosis
<b>Pulmonary fibrosis, other lung damage</b>	According to combination of symptoms, signs, PFT, and confirmed radiologically or by lung biopsy AND/OR persistent cavity, abscess, atelectasis
<b>Pleural fibrosis</b>	Confirmed by radiological investigation.
<b>Pulmonary hypertension</b>	Combination of signs (loud P2) and symptoms confirmed by cardiological investigation.
<b>Pulmonary thrombo-embolic disease/infarction</b>	According to symptoms and signs and confirmed by radiological investigation
<b>Chronic respiratory insufficiency</b>	Chronic hypoxemia requiring oxygen therapy and/or pulmonary function less than 50%, lasting $\geq 3$ months

<b>6. Cardiovascular</b>	
<b>Coronary artery aneurysm or stenosis</b>	Aneurysm (>3-4mm) or stenosis lasting $\geq 3$ months, determined by echocardiography and cardiology review, with or without angioplasty.

<b>Myocardial infarction</b>	Signs and symptoms confirmed by ECG changes and/or cardiac enzyme elevation, lasting $\geq 3$ months
<b>Subsequent myocardial infarction</b>	Another myocardial infarction occurring at least 3 months apart from the previously scored event
<b>Chronic cardiomyopathy</b>	Chronic ventricular dysfunction confirmed by echocardiography and cardiology review, lasting $\geq 3$ months
<b>Valvular heart disease</b>	Significant diastolic or systolic murmur due to valve disease as confirmed by echocardiography, lasting $\geq 3$ months
<b>Pericarditis or pericardiectomy</b>	Symptomatic pericardial inflammation or constriction for at least 3 months or pericardiectomy
<b>Chronic arrhythmia</b>	Significant cardiac rhythm disorder lasting $\geq 3$ months, confirmed by ECG and cardiology review
<b>Major vessel stenosis</b>	e.g. aorta, carotid, brachial, femoral, renal or mesenteric artery stenosis, lasting $\geq 3$ months, documented on Doppler ultrasound or angiography (MR, CT or catheter arteriography)
<b>Absent pulses in one limb</b>	Detected clinically
<b>2<sup>nd</sup> episode of absent pulses in one limb</b>	Detected clinically, at least 3 months apart from the previously scored event and lasting $\geq 3$ months
<b>Minor tissue loss</b>	Finger pulp or similar non-extensive tissue loss.
<b>Major tissue loss</b>	e.g. the loss of phalanx/digit(s) or limb(s), including by surgical resection; tissue loss requiring skin grafting
<b>Subsequent major tissue loss</b>	Another major tissue loss occurring at least 3 months apart from the previously scored event
<b>Chronic claudication of extremities</b>	Localised muscular or extremity pain elicited usually by physical activity present for $\geq 3$ months
<b>Complicated venous thrombosis</b>	Persistent limb swelling with clinical and/or ultrasound evidence of venous stasis, lasting $\geq 3$ months, with or without ulceration

## 7. Abdominal

<b>Chronic mesenteric insufficiency/pancreatitis</b>	Abdominal pain from bowel or pancreatic ischaemia/pancreatitis confirmed on angiography / enzyme changes, lasting $\geq 3$ months
<b>Gut infarction / resection</b>	Infarction or resection of bowel below duodenum; or of gall bladder, spleen, or liver
<b>Oesophageal stricture / surgery</b>	Stricture confirmed radiologically or by endoscopy, lasting $\geq 3$ months, with or without dilatation / surgery
<b>Other GI tract stricture</b>	Stricture confirmed radiologically or by endoscopy, lasting $\geq 3$ months
<b>Chronic peritonitis</b>	Clinical findings confirmed by ultrasound, lasting $\geq 3$ months
<b>Malabsorption / chronic diarrhoea</b>	From small bowel or pancreatic dysfunction requiring nutritional / enzyme supplement to correct or prevent failure to thrive. lasting $\geq 3$ months

## 8. Renal

<b>BP &gt; 95<sup>th</sup> centile or requiring antihypertensives</b>	Systolic or diastolic BP >95th centile for height, age or requiring anti-hypertensive drugs, persisting $\geq 3$ months <a href="http://www.nhlbi.nih.gov/guidelines/hypertension/child_tbl.htm">www.nhlbi.nih.gov/guidelines/hypertension/child_tbl.htm</a>
<b>Proteinuria</b>	Persistent >20 mg protein / mmol creatinine and/or > 0.3 g/24 hours, lasting $\geq 3$ months
<b>GFR 15-60 ml/min/1.73m<sup>2</sup></b>	Estimated or measured, by any locally used method, lasting $\geq 3$ months
<b>End stage renal disease</b>	Chronic kidney disease stage five (GFR < 15 mls/min/1.73m <sup>2</sup> by any

	locally used method), lasting $\geq 3$ months
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<b>9. Neuropsychiatric</b>	
<b>Cognitive dysfunction</b>	Reported impact on school performance or developmental delay (memory deficit, difficulty with calculation, poor concentration, difficulty in spoken or written language), documented on clinical examination (e.g. short mental test score), lasting $\geq 3$ months. Verification required by neuropsychologist / developmental paediatrician for subtle cognitive impairment.
<b>Major psychosis</b>	Severe disturbance of perception of reality characterised by: delusions, hallucinations, incoherence, marked loose associations, impoverished thought content, marked illogical thinking, bizarre, disorganised or catatonic behaviour confirmed as psychosis by psychiatry, lasting $\geq 3$ months.
<b>Seizures</b>	Seizures requiring therapy for $\geq 3$ months.
<b>Stroke/ischaemic lesion</b>	Focal neurological deficit such as paresis, weakness, etc., lasting $\geq 3$ months, plus imaging evidence of lesion OR brain surgery for causes other than malignancy or trauma.
<b>Subsequent stroke</b>	Another stroke occurring at least 3 months apart from the previously scored event
<b>Cord lesion</b>	Lower extremity weakness or sensory loss (usually with a detectable sensory level), usually with loss of sphincter control (rectal & urinary bladder), lasting $\geq 3$ months.
<b>Cranial nerve palsy</b>	Facial nerve palsy, recurrent nerve palsy, oculomotor nerve palsy etc. excluding sensorineural hearing loss and ophthalmic symptoms due to inflammation, lasting $\geq 3$ months
<b>Peripheral neuropathy</b>	Peripheral neuropathy resulting in either motor or sensory dysfunction, lasting $\geq 3$ months.

<b>10. Other</b>				
<b>Primary gonadal failure / delayed puberty</b>	Confirmed by endocrinologist or Ages in parentheses are the approximate 3rd and 97th percentiles for attainment.			
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<b>Secondary gonadal failure</b>	Female: lack of periods for 3 or more months in a girl with established regular menstruation and who is not pregnant or breastfeeding (associated with high FSH). Male: failure or reduced erections and ejaculations (associated with high FSH) or azoospermia
<b>Growth delay / failure to thrive</b>	Empiric clinical assessment or crossing of two centiles on the centile chart (but not when attributed to delay of puberty)
<b>Cushingoid obesity</b>	Weight gain > 10% due to corticosteroid treatment
<b>Marrow failure</b>	Cell-line aplasia, lymphoproliferative disease or myelofibrosis confirmed by bone marrow aspiration and / or haematology opinion
<b>Diabetes</b>	Requiring any type of therapy
<b>Chemical cystitis</b>	Persistent haematuria or shrunken bladder. This does not include acute haemorrhagic cystitis which should be scored as Adverse Drug Reaction.
<b>Malignancy</b>	Documented by histopathologically and either requiring treatment or terminal disease
<b>Other</b>	Any feature considered by patient or doctor to be an important “scar” (residual permanent damage) or consequence which has arisen since onset of disease
<b>School absence</b>	Each week includes 5 school days (ie do not count weekend) Partial day attendance for more than a half day (ie for 3-6 hours) is counted as a full day Any attendance less than half (ie 1-3 hours) is counted as a half day Home teaching is only included as school attendance if it had been the intention despite the patient becoming unwell. If a holiday period, estimate the likely attendance had it been during school terms (considering attendance before and after the holiday break and any hospital attendances during the break)