

The NPC-SI calculator for individuals older than four years

How to use the calculator

Carefully assess the signs and symptoms, selecting those that are present in the individual you wish to screen.
Then calculate the total Risk Prediction Score (RPS), before referring the individual on for further testing as appropriate.

What to do next

Understanding the results

Risk Prediction Score	Referral advice
< 40	LOW likelihood of NPC Discount other possible causes
40-70	MODERATE likelihood of NPC Contact your nearest NPC referral centre to discuss
≥ 70	HIGH likelihood of NPC Refer the individual to an NPC referral centre for immediate testing

Need more information?

If you need further information about the symptoms included in the NPC-SI or how to assess these;
visit npc-si.org

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	Visceral	Neurological	Psychiatric
Very strong 40pts / item		<input type="checkbox"/> Vertical supranuclear gaze palsy <input type="checkbox"/> Gelastic cataplexy	
Strong 20pts / item	<input type="checkbox"/> Prolonged unexplained neonatal jaundice or cholestasis <input type="checkbox"/> Isolated unexplained splenomegaly (historical ± current) with or without hepatomegaly		<input type="checkbox"/> Presenile cognitive decline or dementia
Moderate 10pts / item		<input type="checkbox"/> Ataxia, clumsiness or frequent falls <input type="checkbox"/> Dysarthria and/or dysphagia <input type="checkbox"/> Dystonia	<input type="checkbox"/> Psychotic symptoms
Weak 5pts / item		<input type="checkbox"/> Acquired & progressive spasticity	<input type="checkbox"/> Treatment-resistant psychiatric symptoms <input type="checkbox"/> Other psychiatric disorders
Ancillary 1pts / item	<input type="checkbox"/> Hydrops fetalis <input type="checkbox"/> Siblings with foetal ascites	<input type="checkbox"/> Hypotonia <input type="checkbox"/> Delayed developmental milestones <input type="checkbox"/> Seizure (partial or generalised) <input type="checkbox"/> Myoclonus	<input type="checkbox"/> Disruptive or aggressive behaviour in adolescence and childhood
Category total	Total points <input type="text"/>	Total points <input type="text"/>	Total points <input type="text"/>
Add additional points if symptoms across more than one category	Visceral & Neurological symptoms (40pts)		<input type="text"/>
	Visceral & Psychiatric symptoms (40pts)		<input type="text"/>
	Neurological & Psychiatric symptoms (20pts)		<input type="text"/>
Family history 40 pts / 1.degree 10 pts / 2.degree	<input type="checkbox"/> Parents or siblings with NP-C <input type="checkbox"/> Cousins with NP-C		<input type="text"/>

Final score