

The NPC-SI calculator for individuals under 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
<3	LOW likelihood of NPC Discount other possible causes
3-5	MODERATE likelihood of NPC Contact your nearest NPC referral centre to discuss
≥6	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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	CNS	Spleen	Liver	Lung	Natal
4pts / Cardinal sign		<input type="checkbox"/> Splenomegaly		<input type="checkbox"/> Pulmonary infiltrates	
3pts / Cardinal sign	<input type="checkbox"/> Cataplexy		<input type="checkbox"/> Prolonged jaundice		
2pts / Cardinal sign	<input type="checkbox"/> Vertical supranuclear gaze palsy		<input type="checkbox"/> Direct bilirubinemia		<input type="checkbox"/> Foetal oedema or ascites
1pt / Cardinal sign			<input type="checkbox"/> Hepatomegaly		
Modifiers: Add if presence of...	↓ CNS signs	↓ Splenomegaly			
2pts / modifier	<input type="checkbox"/> Ataxia*				
1pt / modifier	<input type="checkbox"/> Cognitive decline	<input type="checkbox"/> Ataxia* <input type="checkbox"/> Cognitive decline			
Category Total	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>

Family history

6pts	<input type="checkbox"/> Parents or siblings with NP-C <input type="text"/>
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*Ataxia may present as gross motor clumsiness in young children

Final score