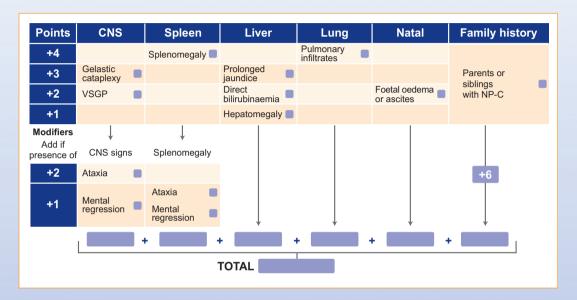# Calculating a risk prediction score

1. Work your way through the disease categories adding points as appropriate
2. Pay close attention to the co-occurrence of ataxia and/or mental regression with central nervous system (CNS) or spleen symptoms
3. Calculate the total RPS for your patient by adding the totals for each category
4. Refer your patient as appropriate

Points	CNS	Spleen	Liver	Lung	Natal	Family history
+4		Splenomegaly ☐		Pulmonary infiltrates ☐		
+3	Gelastic cataplexy ☐		Prolonged jaundice ☐			Parents or siblings with NP-C ☐
+2	VSGP ☐		Direct bilirubinaemia ☐		Foetal oedema or ascites ☐	
+1			Hepatomegaly ☐			

Modifiers
Add if presence of

	CNS signs	Splenomegaly
+2	Ataxia ☐	
+1	Mental regression ☐	Ataxia ☐ / Mental regression ☐

+6 (Family history)

◻ + ◻ + ◻ + ◻ + ◻ + ◻

TOTAL ◻

Interpreting the results

The **NP-C SI 0–4y** is derived from an independent patient cohort; the total infantile RPS therefore cannot be directly compared with RPS derived from other NP-C SI tools

Total RPS	Suspicion of NP-C	Action
<3	Low likelihood	Ensure other possible causes have been discounted
3–5	Moderate likelihood	Contact your nearest NP-C referral centre for further discussion
≥6	High likelihood	Refer patient to an NP-C centre for immediate testing

NPC-SI.com
Niemann-Pick Type C Suspicion Index