CANADIAN PNH NETWORK
Partners in Care for PNH

Follow-up of PNH patients after initial workup – routine visit (every 3-4 months) for all patients, regardless of treatment
Frequency determined by treatment, disease severity and local support

History and physical exam	Rationale for test/evaluation
Monitor the "7 P's": **1.** Pep: rating fatigue **2.** Paroxysms: have there been any episodes of increased hemolysis or of hemoglobinuria **3.** Pallor: anemia, transfusions **4.** Pulmonary: dyspnea **5.** Pain: esophageal spasm, chest/abdominal pain, need for narcotic analgesia **6.** Penis: erectile dysfunction (if applicable) **7.** Pregnancy: assess possibility of or plans of pregnancy	• Physical symptoms will help determine management strategy

Hematology	
1. FLAER/RBC flow **2.** CBC **3.** Reticulocytes **4.** LDH **5.** DAT **6.** D-dimer **7.** Serum ferritin	• Flow cytometry required to monitor any expansion of the PNH clone • CBC to track anemia and other cytopenias • Reticulocytes and LDH to detect active hemolysis • DAT to confirm that hemolysis is not autoimmune • D-dimer to assess thrombotic risk • Serum ferritin to assess potential iron overload or deficiency – if normal, it can be tested less frequently (e.g., every 6 months) • Assess for additional/alternative causes of anemia (e.g., bleeding, iron deficiency, vitamin B12)

Renal	
1. Electrolytes **2.** Creatinine, estimated CrCl **3.** Microalbumin **4.** Urinalysis (routine & microscopic)	• Important to compare markers of renal function across visits to assess any deterioration

Other	
1. BNP **2.** Previous anti-complement therapy treatment history (if any) and outcomes **3.** Imaging for thromboembolic events	• If BNP is stable/normal, ongoing monitoring can be less frequent (e.g., annually) • Low clinical threshold for dedicated imaging to rule out arterial or venous thrombosis based on clinical history

RBC = red blood cell; CBC = complete blood count; LDH = lactate dehydrogenase; DAT = direct antiglobulin test; CrCl = creatinine clearance; BNP = B-type natriuretic peptide

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Follow-up – additional evaluations at routine visit for patients on anti-complement therapy

History and physical exam	Rationale for test/evaluation
Additional "2 P's": **1.** Pyrexia: history of fever/infections **2.** Prophylaxis: history of penicillin or other antibiotics for meningococcal prophylaxis, requirement for new/renewed prescription	• Patients on anti-complement therapy should report any infections/fevers they experience • Prophylaxis is recommended with anti-complement therapy treatment even if patient is vaccinated

Additional evaluations – to be done at least once in patients on anti-complement therapy

Other

1. Anti-meningococcal titres (once available)	• Anti-meningococcal titres coming soon to Canada; will help assess whether prior vaccination provided effective protection

Additional evaluations – annual visit (all patients, whether on anti-complement therapy or not)

Other

1. 2-D echocardiogram **2.** Viral serology in transfused patients	• Monitor cardiac function and assess for pulmonary hypertension

Additional evaluations – patients with small/asymptomatic clone

Hematology

1. Evaluate clone size every 6-12 months	• Monitor any changes in clone size and how they correlate to development of symptoms