

Table 6. Recommendations for transfusion support

GENERAL PRINCIPLES	
Indications and timing	Therapeutic considerations
<ul style="list-style-type: none"> Any patient with severe anemia Patient within 12 months of life Patient not responding to steroids or experiencing significant side effects Patient responding to steroids and experiencing acute Hb drop (e.g., due to viral illness) Patient on steroid holiday (to improve growth during adolescence) Pregnant patient with anemia 	<p>General:</p> <ul style="list-style-type: none"> Hepatitis B vaccination RBC antigen typing and repeat RBC antibody screening <p>Hb goal prior transfusion (nadir Hb):</p> <ul style="list-style-type: none"> ≥9-10g/dL or a higher level at which the patient is asymptomatic, independent of age <p>Transfusion process:</p> <ul style="list-style-type: none"> ¹Volume: 10-15ml/kg (children), ~2-3 RBC units (adults) ²Interval: every 3 (2-4) weeks
ADVERSE EFFECTS AND CLINICAL PROBLEMS	
Iron overload	Start early chelation (Table 8)
Clinically significant anemia, especially days before transfusion	Increase transfusion volume or decrease transfusion interval, “catch-up” transfusion
Blood-transmitted pathogens	Hepatitis B vaccine Virus testing (HIV, Hepatitis B and C) at least yearly
Higher transfusion requirements	Rule out alloimmunization and hypersplenism (rare in DBA syndrome), hemorrhage

¹ Higher transfusion volumes occasionally required across all ages (i.e., ~20ml/kg).

² Interval may be longer in patients with some erythropoiesis who maintain Hb ≥9-10g/dL for longer period of time.