Thomas AE, Halsey C (eds): Controversies in Pediatric and Adolescent Hematology.
Pediatr Adolesc Med. Basel, Karger, 2014, vol 17, pp 42–66 (DOI: 10.1159/000350346)

Hematological Problems in Pediatric Intensive Care

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Abstract

Hematologic abnormalities are common in children admitted to pediatric intensive care units (PICUs). Some are primary and related to the underlying hematologic disease in the child such as severe anemia in a young child with sickle cell disease and an acute splenic sequestration crisis. Others are secondary to other underlying conditions such as disseminated intravascular coagulation secondary to sepsis. Perhaps the most common association is with surgery or following trauma. The approach to the diagnosis and management of hematologic disorders in children cared for in PICUs requires an appreciation of disorders, some very rare, where delays in diagnosis and initiation of effective treatment can be associated with significant morbidity/mortality (e.g. thrombotic thrombocytopenic purpura, hemophagocytic lymphohistiocytosis) as well as more 'mundane' but equally important decisions such as transfusion triggers for replacement of packed red blood cells, fresh-frozen plasma, cryoprecipitate or platelets. In all situations, a best practice approach based on available published evidence and expert opinion is recommended. In large tertiary/quaternary care centers, a close working relationship between the pediatric intensivist staff and the clinical/laboratory hematology services is essential for optimizing the care of critically ill children with hematologic problems cared for in PICUs.

A wide range of hematologic problems occur in patients admitted to pediatric intensive care units (PICUs). These may reflect primary hematologic disorders or arise as complications of other underlying conditions. The aim of this chapter is to focus on hematologic disorders seen in children aged >4 months to <18 years of age

admitted to PICUs with an emphasis on conditions that are common or present challenges in diagnosis or management to the pediatric intensivist. For problems in neonates and younger infants (ages 0–4 months), the reader should consult other sources.

Disorders of Red Blood Cells

Anemia

Anemia is a condition in which the hemoglobin level, as a surrogate marker for the circulating red blood cell (RBC) mass, is below normal for age. An important caveat, particularly relevant to the ICU setting, is that in situations of clinically significant acute blood loss the correlation between the circulating hemoglobin level and the circulating RBC mass is often poor. In such situations the decision to transfuse donor RBCs should take into consideration other clinical parameters such as the cardiorespiratory status of the patient and other factors affecting tissue oxygenation.

Bone marrow failure (BMF), hemolysis and blood loss are the three main mechanisms that lead to anemia severe enough to need admission to the PICU. Children with severe acute or chronic anemia leading to congestive heart failure, disseminated intravascular coagulation (DIC), those with complications of inherited or acquired severe BMF states and children with sickle cell disease (SCD) presenting with certain crises (e.g. stroke, acute splenic sequestration and chest crises) are frequently managed in the PICU. The clinical findings and management of such patients are dependent on the rate at which the anemia develops, the capacity of the cardiopulmonary system to compensate and the underlying disorder or disorders that resulted in anemia.

Approximately one-half of children admitted to PICUs with anemia will receive RBC transfusions, most within the first 48 h, reflecting events such as blood loss associated with surgery and/or trauma preceding the admission [1]. Importantly, after the first 48 h, iatrogenic blood loss for laboratory testing is a significant contributor to the need for RBC transfusion in PICU, especially in young children <10 kg body weight.

There are well documented complications associated with RBC transfusions, including transfusion-transmitted infections, transfusion-related acute lung injury, transfusion-associated circulatory overload, acute hemolysis, allergic reactions and transfusion-induced immune modulation. Given the frequency of anemia in children admitted to PICUs and the potential adverse effects of RBC transfusions, a decision to transfuse RBCs should be based on the individual patient characteristics rather than a predetermined hemoglobin level [2, 3] (table 1a). Despite the very low risk of transmitting HIV or hepatitis C infection (the absolute risk may be different between countries), recent emphasis is appropriately directed toward minimizing the amount of RBC transfusions in PICU patients.

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Table 1. Use of blood products in children admitted to PICU (see more details in the text)

a RBC transfusion

Key points

Suggested transfusion 'triggers'

- A hemoglobin transfusion threshold of 70 g/l can decrease RBC transfusion requirement without an increase in adverse outcome

Strategies to reduce RBC transfusion requirements

- Limit blood sampling for laboratory testing to that essential for the wellbeing of the child

RBC transfusion in children with SCD

- Simple or exchange RBC transfusions for arterial ischemic stroke, acute chest syndrome, multiorgan failure, splenic sequestration, perioperative, aplastic crisis
- Exchange (not simple) transfusion is recommended if the baseline hemoglobin level is >90 g/l and/or if a rapid decrease in HbS level to below 30% is required

Pediatric dose: 10–15 ml/kg of packed RBCs (10 ml/kg for units stored in CPDA-1, otherwise, 15 ml/kg for AS-1, AS-3, AS-5, SAGM units). In children >20 kg body weight the transfusion volume is rounded to the nearest number of packed RBC units.

b Granulocyte transfusion

Key points

Granulocyte transfusions are rarely used in children

Clinical efficacy of granulocyte transfusion is still uncertain/controversial

Hematology/transfusion medicine consultation is recommended if granulocyte transfusion is being considered in children with severe neutropenia who satisfy all of the following criteria:

- Very severe neutropenia (absolute neutrophil counts $<0.2 \times 10^9$/l) due to congenital or acquired bone marrow failure unresponsive to G-CSF therapy
- Proven or highly probable fungal or bacterial infection unresponsive to appropriate anti-infective therapy
- Neutrophil recovery expected within the near future and/or definitive or curative therapy (e.g. hematopoietic stem cell transplantation) is planned

Pediatric dose: 'buffy coat' preparation 10–20 ml/kg or apheresis granulocyte concentrate – 1 unit. All granulocyte preparations must be irradiated before transfusion.

c Platelet transfusion

Key points

Suggested prophylactic platelet transfusion 'triggers' for thrombocytopenia due to production failure

- In most cases a threshold of 10×10^9/l is safe and cost-effective
- Threshold of 20×10^9/l in neonates, in children with sepsis, and children with a higher bleeding risk (e.g. children with underlying bleeding disorders)
- Threshold of 30×10^9/l for children with brain tumors
- Threshold of 50×10^9/l for children requiring lumbar puncture, surgery
- Threshold of 100×10^9/l for children requiring neurosurgical procedures

Platelet refractoriness

- Defined as an inadequate rise in platelet counts as measured within 10 min to 1 h of a transfusion of an adequate number of platelets

Pediatric dose: 10–15 ml/kg or 1 U whole blood-derived platelets/per 10 kg body weight, up to the equivalent of 1 U of apheresis platelet or 5–6 U of whole blood-derived platelets.

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