

Epidural Anesthesia in Diamond-Blackfan Anemia: Hematologic and Cardiac Perspectives

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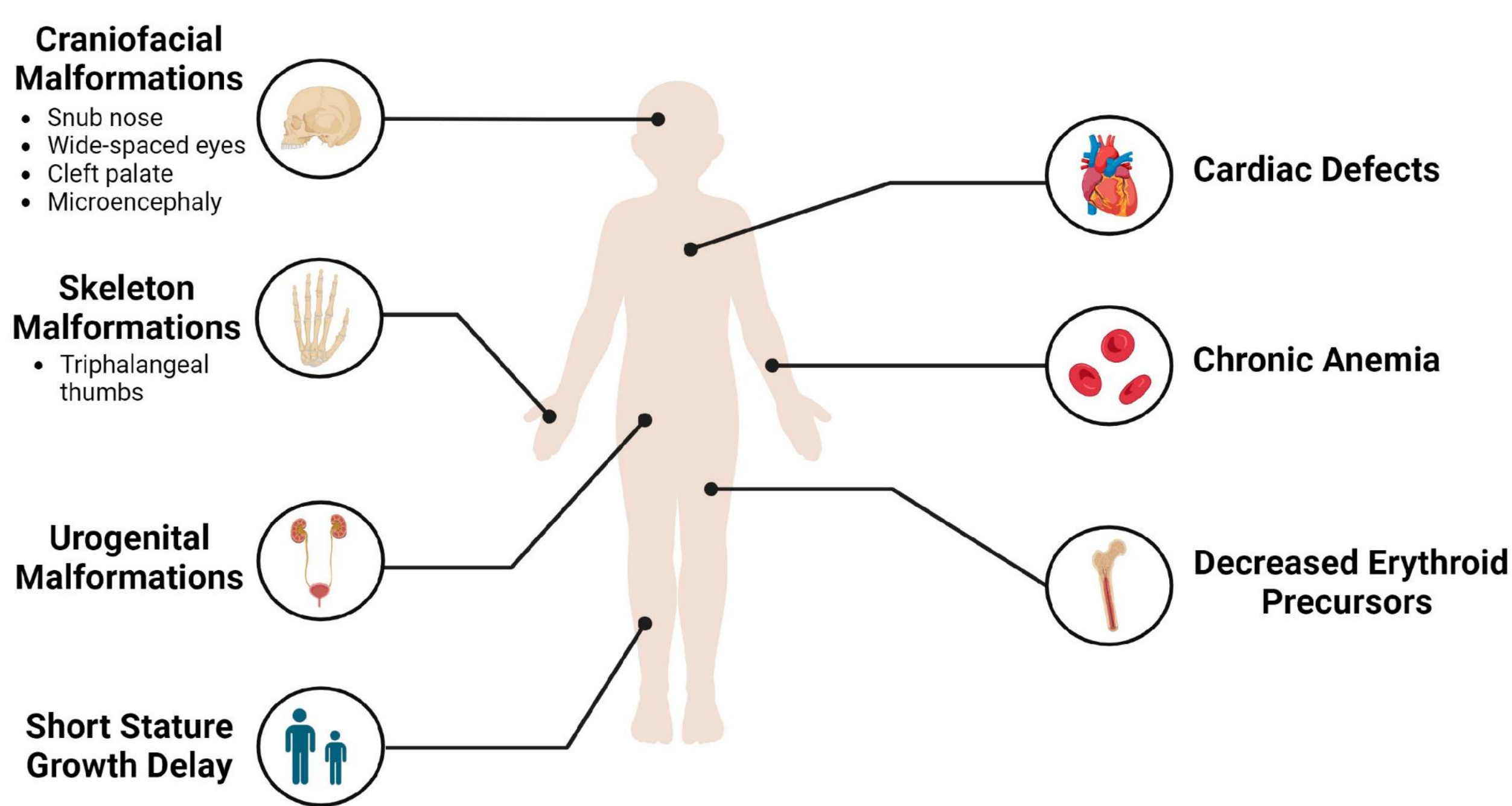
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Background

Diamond–Blackfan anemia is a rare congenital bone marrow failure syndrome marked by severe macrocytic anemia due to reduced red blood cell production. It is caused by mutations in ribosomal protein genes and may be associated with congenital anomalies, though many patients appear otherwise normal. Management often involves chronic transfusions and/or steroids, with iron overload being a common complication that can lead to cardiac, liver, and endocrine dysfunction.



Patient Presentation

43 y/o G1P0000 Female at 34w5d presenting for induction of labor in the setting of preeclampsia w/ severe features
PMHx:

- Diamond-Blackfan Anemia complicated by iron overload from chronic infusions and thrombocytopenia from bone marrow suppression
- Restrictive cardiomyopathy w/ diastolic heart failure

Anesthetic Plan

Primary plan: Epidural analgesia for induction of labor

Special Considerations

- Maintain Hgb>10g/dL with transfusions
- If retained placenta-> extend epidural for surgical anesthesia
- Postpartum hemorrhage-> prepare uterotonics such as Pitocin and carboprost

Goals: To maintain normovolemia, monitor for fluid overload. Avoid anemia and be prepared for possible bleeding. Minimize hemodynamic shifts to prevent complications.

Clinical Course

The patient underwent a forceps-assisted vaginal delivery with epidural anesthesia. Delivery was complicated by a retained placenta, necessitating a dilation and curettage. She then developed a postpartum hemorrhage, which was managed with transfusion of 3 units of packed red blood cells to correct acute blood loss anemia. Uterotonic therapy with carboprost was administered to improve uterine tone and control ongoing bleeding. Hemodynamic stability was achieved following these interventions. Postoperatively, the patient developed pulmonary edema, likely multifactorial in the setting of underlying restrictive cardiomyopathy from transfusional iron overload and the intravascular volume shifts associated with delivery and transfusion. She required supplemental oxygen via 4 liters nasal cannula for hypoxia and was treated with intravenous diuretics, resulting in improvement of respiratory status. Her symptoms resolved fully prior to discharge, with stable oxygenation and no further evidence of fluid overload.

Discussion

When considering epidural analgesia in a patient with Diamond-Blackfan Anemia (DBA), several factors must be addressed to ensure patient safety. Postpartum hemorrhage should be anticipated in severely anemic patients. Having packed red blood cells available prior to delivery allowed for rapid hemoglobin correction. Although platelet counts are typically normal in DBA, chronic transfusions and pre-eclampsia can result in thrombocytopenia. A CBC should be obtained within six hours prior to epidural placement, as platelet counts below 70,000-80,000/ μ L increase the risk of spinal hematoma. Chronic transfusions can also lead to restrictive cardiomyopathy, resulting in pre-load dependent cardiac output. Epidural-induced vasodilation can decrease preload, increasing the risk of hypotension, cardiac collapse, and pulmonary edema. To minimize hemodynamic shifts, a slowly titrated epidural infusion with low dose anesthetics, such as 0.0625% bupivacaine+fentanyl 2mcg, is recommended. Though the patient required treatment for postoperative complications, the ability to avoid catastrophic events illustrates the importance of carefully tailored anesthetic planning in patients with significant comorbidities.

References

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