

in children with sickle cell disease.

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Abstract:

Despite the high frequency of sickle cell disease in Europe, the disease is poorly managed. Critical periods are the hospital stays during which the anaesthesiologist plays an important role. Understanding the molecular basis of polymerization processes of haemoglobin S can help to avoid triggering a crisis. Differentiation of the various haemoglobin phenotypes helps to estimate the individual perioperative risk. Knowledge of the patient's history and the actual haemoglobin S level facilitates general anaesthesia, surgery and postoperative care. Damage to liver, spleen, eyes, bones, lung and central nervous system increases the perioperative risk. Preoperative preparation includes early admission, intravenous volume substitution, continuing pain therapy and prophylactic antibiotic medication. General anaesthesia seems to be better for patients with a high-risk profile rather than regional anaesthesia. Careful perioperative and postoperative monitoring should allow hypoxaemia, hypovolaemia, hypothermia, acidosis and overtransfusion to be avoided. Effective pain therapy includes a combination of opioids with peripherally acting analgesia.