Co-management Guide

Pediatric Hematology/Oncology	Sickle Cell Disease Vaso Occlusive Crisis (VOC)

Guidelines Referenced	DeBaun, M., Vichinsky, E. P. (2019). Evaluation of acute pain in sickle cell disease. *UpToDate*. https://www.uptodate.com/contents/evaluation-of-acute-pain-in-sickle-cell-disease?search=sickle%20cell%20disease&topicRef=7144&source=see_link DeBaun, M., Vichinsky, E. P. (2019). Vaso-occlusive pain management in sickle cell disease. *UpToDate*. https://www.uptodate.com/contents/vaso-occlusive-pain-management-in-sickle-cell-disease?search=sickle%20cell%20disease&topicRef=7114&source=see_link#H530226134 Yawn, B., Buchanan, G., Afenyi-Annan, A., Ballas, S., Hassell, K., James, A.,…John-Sowah, J. (2014). Management of sickle cell disease: Summary of the 2014 evidence-based report by expert panel members. *Journal of American Medical Association*, *312*(10):1033-1048. doi:10.1001/jama.2014.10517
Background	Vaso Occlusive Crisis (VOC) or acute pain crisis is the most common complication associated with Sickle Cell Disease (SCD). VOC is usually accompanied by severe, debilitating pain that requires prompt intervention. Almost all patients with SCD will experience VOC within their lifetime, and episodes can occur in patients as young as 6 months of age. The most common sites of pain in VOC are back, chest, abdomen, long bones, hands and feet. Urgent management is important in cases of VOC to prevent the development of further complications such as acute chest syndrome (ACS). Other potentially life threating complications/comorbidities must also be excluded when evaluating pain in SCD.
Initial Evaluation	Elicit a detailed pain history (location, timing, duration, frequency, radiation, associated symptoms, alleviating and exacerbating factors). Have patient describe quality and severity of pain using age appropriate pain scale. Obtain full set of vital signs including o2 sat. Perform physical exam of affected area (ie musculoskeletal exam, abdominal exam, etc..). Palpate for splenomegaly. Perform respiratory assessment (auscultate lung fields, assess quality of breathing, perfusion/skin tone). Assess for any neurological deficits (ie mental status changes, headaches, neurologic exam).

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Initial Management	Manage mild to moderate pain with ibuprofen 10mg/kg/PO Q6HPRN and severe pain per with either ibuprofen 10mg/kg PO + Tylenol 15mg/kg PO or with opioid such as oxycodone 0.1mg/kg PO or hydrocodone with Tylenol (0.1-0.15mg/kg of hydrocodone) PO x 1. Alternative pain relief measures include PO hydration, warmth, massage, distraction. DO NOT TREAT WITH ICE/COLD THERAPY If pain is relieved by above interventions, continue to manage at home while promoting rest, hydration, warmth and other beneficial alternative pain relief techniques (as per pain mgmt. guideline). If available through your office, provide IVF bolus 20mL/kg over 1 hour. If pain persists ~45 minutes after above interventions or worsens, consult Pediatric Hematology/Oncology (PHO).
When to Refer	Notify PHO: If patient is febrile (temp 101 deg F or greater). Obtain blood cultures and notify PHO. With confirmed abnormal vital signs including hypoxia If pain fails to improve after treatment with opioid analgesic or if pain fails to improve with pain treatments available through your office, notify PHO. If patient or family reports that the pain is abnormal for this patient (significantly different from previous pain episodes by location, severity or other characteristics) If pain associated with extremity swelling or deformity unstable vital signs, If abdominal distention/ splenomegaly present With abnormal respiratory exam findings, notify PHO. With mental status changes and/or severe headaches If Hgb significantly increased or decreased from baseline If Low reticulocyte count

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	If severe pain with recent blood transfusion If creatinine is elevated from baseline Significantly worsened icterus/ or darkened urine Persistent limp that fails to improve with available pain mgmt interventions	
Pre-Visit Work Up	Full set of vital signs and physical exam as above in initial evaluation Labs: CBC c diff, CMP, retic, bili panel. Blood culture if febrile (temp 101 degrees F or greater) Chest xray PA/Lat for abnormal respiratory exam in stable patient Abdominal ultrasound if splenomegaly palpated in stable patient	
Co-management Strategy (as appropriate)	**Specialist scope of care** Management of sickle cell complications from VOC or causing VOC (as above) Management of VOC that requires hospitalization, IV narcotics	**Primary care scope of care** Management of VOC that can be treated with PO meds, complementary therapy and IVF if available
Return to Primary Care Endpoint	Pain improving on PO pain medication, clinically stable	