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MANAGEMENT OF ACUTE CHEST SYNDROME IN THE CHILD WITH SICKLE CELL DISEASE

DEFINITION: An acute illness associated with lower respiratory symptoms, hypoxemia, or new infiltrate on CXR.

CONSULTS: Hematology
Consider Pulmonology

MONITORING:
1. Hospitalize.
2. Vital signs with BP q 2-4 hr.
3. Pain intensity rating at least q 4 hr.
4. Continuous pulse ox.
5. Record I+O, daily weight.
6. Consider transfer to ICU for severe or rapidly progressive illness.

DIAGNOSTICS:
1. CBC, diff, platelet count, and retic count initially and daily until improving. Compare with patient's baseline values.
2. CXR initially, repeat for significant changes or clinical deterioration.
3. Consider:
   - Type and crossmatch for severe illness or if Hb <6 gm/dl or 15% or more below baseline. Request leukocyte-depleted and, if available, C, E, Kell-compatible (requires minor antigen phenotype) and sickle-negative RBC. In absence of alloantibodies, urgent transfusion should not be delayed by search for minor-antigen matched units.
   - Blood cultures if febrile or history of recent fever.
   - Consider blood gas or arterial line for severe illness.
   - Renal (BUN, creat) and liver (fractionated bili, ALT) function tests for severe illness or if diffuse encephalopathy present (R/O acute multiorgan failure syndrome).

FLUIDS, NUTRITION, GENERAL CARE:
1. Maintain "euvolemia." IV (D_5 ¼) + P.O. @ 1x maintenance. More fluid is appropriate only if patient is dehydrated or if insensible losses are increased (e.g. persistent fever).
2. Incentive spirometry - 10 breaths q 2 h when awake. Consider soap bubbles or pinwheels for younger patients.
3. Encourage ambulation, activity.

MEDICATIONS/TREATMENTS:
1. Oxygen to pulse ox ≥ 92% or ≥ baseline value, if baseline >92%.
2. Albuterol nebs q 4-6 hr, especially if patient has history of reactive airway disease or wheezing on exam. Consider CPT or vibratory vest. Continue any outpatient asthma meds (e.g., Singulair, Advair) if applicable.
3. Consider positive pressure ventilation (nasal CPAP or mask BiPAP) for patients with poor respiratory effort or reduced ventilation. Consider transfer to ICU for BIPAP > 12/6 mmHg or > 50% O_2.
4. Consider red cell transfusion:
   - Simple transfusion for moderately severe illness, especially if Hb >1 gm/dl below baseline (do not transfuse acutely to Hb >10 gm/dl, Hct >30%).
   - Partial exchange transfusion to Hb 10 gm/dl and Hb S or Hb S+C (patient's RBC) ≤ 30% for severe or rapidly progressive disease (may require transfer to ICU and transfusion medicine consult for erythrocytapheresis). Remove femoral or central venous catheters as soon as possible after exchange transfusion to reduce risk of thrombosis.
5. Ceftriaxone 50-75 mg/kg IV q 24 hr (2 gm max single dose) or cefotaxime 50 mg/kg IV q 8 h (2 gm max single dose). Substitute meropenem 20 mg/kg IV q 8 h (1 gm max single dose) for patient with known or suspected cephalosporin allergy. Prophylactic penicillin should be discontinued while patient is receiving broad-spectrum antibiotics.
6. Azithromycin 10 mg/kg po first dose (500 mg max single dose), then 5 mg/kg qd (250 mg max single dose), or other macrolide antibiotic.
7. Strongly consider adding vancomycin 15-20 mg/kg IV q 8 hr (1 gm max single dose) for severe illness or for suspected *S. aureus* infection. Draw peak and trough vancomycin levels after 3rd or 4th dose if vancomycin to be continued > 48 hr.

8. Opioid
    - Morphine sulfate 0.05 - 0.15 mg/kg/dose IV q 2 hr or 0.05 - 0.1 mg/kg/hr continuous infusion or via PCA. (For PCA give 1/2-2/3 of total maximum dose by continuous infusion, with 1/3-1/2 via PCA boluses.) Total morphine dose, continuous infusion plus boluses, above 0.1 mg/kg/hr may be required, especially for opioid-tolerant patients, but should be used with caution.
    - Nalbuphine (Nubain) 0.3 mg/kg IV q 3 hr, 0.2 mg/kg IV q 2 hr, or 0.1 mg/kg/hr continuous infusion. Do not use nalbuphine for patients receiving chronic opioids (e.g. MS Contin, Oxycontin).
    - Other opioids such as hydromorphone (Dilaudid) 0.015-0.02 mg/kg IV q 3-4 hr or fentanyl may be appropriate in selected cases. Repeated doses of meperidine (Demerol) should be avoided because of the risk of seizures.
9. NSAID
    - Ketorolac (Toradol) 0.5 mg/kg (30 mg maximum dose) IV q 6 hr or ibuprofen 10 mg/kg po q 6-8 hr if no contraindication present (i.e. gastritis, ulcer, dehydration, coagulopathy, or renal impairment). Limit Ketorolac and more frequent dosing of ibuprofen to 5 days per month maximum duration.
10. Consider one dose of furosemide 0.5 mg/kg IV if signs of fluid overload present.
11. Acetaminophen 10-15 mg/kg po q 4 hr or prn T >38.0^{o}C (75 mg/kg/day or 4 gm/day max).
12. Consider ducosate and/or laxative for opioid-induced constipation.
13. Consider diphenhydramine (0.5 mg/kg po q6h, 50 mg/dose max), hydroxyzine (0.5 mg/kg po q6h, 50 mg/dose max), or low-dose nalbuphine (10-20 mcg/kg IV q6h) prn pruritis. Offer menthylated lotion prn pruritis.
14. Consider promethazine (0.25-0.5 mg/kg po q6h, 25 mg/dose max) or ranitidine (2 mg/kg po q12h, 150 mg/dose max) prn nausea.
15. Offer heating pads or other comfort measures previously used by patient. Avoid ice or cold packs.
16. Reassess pain control on a regular basis (at least twice daily and after any change in analgesics) by using age-appropriate pain scale and by discussing efficacy and side effects with patient/family. Analgesics may be weaned as tolerated by decreasing dose, <u>not</u> by prolonging interval between doses. Discuss analgesic changes with patient/family.
17. See other Clinical Guidelines for acute splenic sequestration, aplastic crisis, stroke, priapism, if present.

<u>DISCHARGE CRITERIA</u>:

1. Improved pulmonary symptoms and documentation of adequate oxygenation on room air.
2. Afebrile ≥ 24 hr. and negative cultures for ≥ 24-48 hr if applicable.
3. Stable hemoglobin/hematocrit.
4. Taking adequate oral fluids and able to take po medications if applicable.
5. Adequate pain relief, if needed, with oral analgesics.
6. Follow-up plans coordinated with hematology service. On a case by case basis, consider follow-up pulmonary function testing and the possibility of chronic transfusions or hydroxyurea.

These guidelines do not indicate an exclusive course of treatment or serve as a standard of care. Variations based on a physician's best medical judgement may be appropriate in individual cases.