

Post-operative management after Kasai

Goals

1. To establish a care guide for the management of infants following Kasai hepatoportoenterostomy for biliary atresia.
2. To tailor therapy based on biological factors that may influence the response to complementary medical therapy after surgery

Rationale

Overall 50% of our infants resume bile drainage after Kasai without supplementary medical treatment. With increased number of reports that identify factors that may influence treatment response (such as age, CMV, and inflammatory signature in the liver) we seek to modify our protocol to take into account patient-specific factors that may improve long-term outcome.

Antibiotics

- Cefoxitin IV x 3-4 days. If patient does well (i.e., colored stools, decrease in bilirubin), switch to treatment dose of Augmentin to complete a 10 day course
- Alternative based on the Juntendo University protocol: Cefoxitin + aminoglycoside
- When completing antibiotic treatment, switch to prophylactic TMP/SMZ x 12 months post-op

Corticosteroids

a) For patients <45 days:

-If the patient restores normal stool color post-operatively, s/he will receive standard post-op care and will be discharged home on fat-soluble vitamins, urso, and antibiotic prophylaxis

-If the patient's stools remain acholic or have color inconsistently (acholic stools are shown on the color card included in the last page: #1, or #2, or #3, with #3 being marginally acholic), s/he will receive a cycle of corticosteroid (see below)

-The transplant hepatology fellow will follow the patients with the surgical team for training purpose, to facilitate communication with hepatology service, and plan for transition to outpatient care

-If the patient receives the steroid cycle, use IV antibiotic in the start of the steroid cycle and then switch to treatment doses of Augmentin x 2 weeks

b) For patients >45 days:

-As for above, standard care if normal stool color is restored

-If the patient's stools remain acholic or have color inconsistently (from stool color card on the last page: #1, or #2, or #3), the liver histology will be reviewed to determine the presence of inflammation. Patients are not to be on Aquadek early post-Kasai so that the stool color can be monitored.

If inflammation is present, s/he will receive a cycle of corticosteroid (see below)

Corticosteroid cycle

The regimen below is designed to provide anti-inflammatory and choleretic effects, with substantial steroid exposure for 2 weeks, followed by 2 weeks of weaning. It also aims at preventing over-exposure to steroids as new cycles are contemplated based on the level of biliary drainage after the initial cycle

- Day 1 - Methylprednisolone 5 mg/kg/day
- Day 2 - Methylprednisolone 4 mg/kg/day
- Day 3 - Methylprednisolone or prednisolone 3 mg/kg/day
- Day 4 - Methylprednisolone or prednisolone 2 mg/kg/day

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- Day 5 - Methylprednisolone or prednisolone 1 mg/kg/day
- Continue prednisolone 1 mg/kg/day x 1 wk
- Wean:
 - 0.75 mg/kg/day x 1 wk
 - 0.50 mg/kg/day x 1 wk
 - 0.25 mg/kg/day x 1 wk
 - Discontinue steroid

Laboratory

- CMP+GGT+Direct bili at day 3-5
- CBC

While on steroids

- Antifungal prophylaxis: Nystatin PO
- Anti-acid (Ranitidine or PPI)

Discharge

- If standard treatment: FU in 2 weeks with surgeon and hepatologist
- If on corticosteroid:
 - Corticosteroid doses per protocol
 - Antifungal prophylaxis: Nystatin PO
 - Augmentin: Treatment doses x 2 weeks
 - TMP/SMZ or another antibiotic: To start after the course of Augmentin
 - Anti-acid (Ranitidine or PPI)
 - No ADEK
 - Urso

Note on CMV-associated biliary atresia:

A clinical report from King's College suggest that the use of ganciclovir post-operatively in infants with biliary atresia improves the rate of bilirubin clearance after HPR. They continue ganciclovir until the serum PCR becomes undetectable. In their open-labeled trial, a few patients also received steroids – without the description of undesired clinical consequences. These patients were started on steroids after HPE but before the test for CMV was available.

