October 23, 2018

Splenic Sequestration and Splenectomy in Sickle Cell Disease

Melisa Ruiz-Gutierrez, MD PhD

Boston Medical Center, Boston, MA

Splenic Sequestration

- Acute splenic sequestration remains a leading cause of death in children with SCD
- Definition: drop in Hg >2g/dL, increased reticulocyte count, acutely enlarging spleen
- Acute splenic sequestration age range: 3 months to 5yrs of age
- Usually associated with viral or bacterial infections
- Clinical symptoms: sudden weakness, pallor, tachycardia, tachypnea and abdominal fullness
- Early recognition of symptoms and prompt treatment improve survival
- Recurrence is common: 50%

Splenic Sequestration by SCD type

- HgSS : most common (90%), age range 3 months to 5 years
  - *Lifetime prevalence 7 – 30%*
  - *Teaching parents to palpate spleen and recognize signs and symptoms*
- HgSC: less common, age range 9 years to adulthood
- HgS/β+: less common, adulthood

Treatment of Acute Splenic Sequestration

- Emergent transfusion of PRBC (best match available, do not delay transfusion)
- Management is geared towards hemodynamic stabilization of patient
- PRBC transfusion will release RBCs sequestered in the spleen, leading to higher Hg/HCT values than predicted from transfused volume
- Spleen will decrease in size after PRBC transfusion

Management After Acute Splenic Sequestration

- Observation
- Chronic Transfusions
- **Splenectomy***

Indication for which approach to take is not clearly defined.

- Observation does not remove risk of fatal episode
- Chronic transfusion risks future sequestration by maintaining spleen function
- Risk of infection after splenectomy for young age
- Total vs partial splenectomy

Observation

- Risk of recurrence is high
- Does not decrease risk of potentially fatal episode

Chronic Transfusions

- Chronic transfusions will decrease risk of splenic sequestration, but can still occur.
- Chronic transfusions may temporarily reverse splenic dysfunction
- Often used in youngest population to allow growth to age where splenectomy is safer (3-5yrs).
- Concern that chronic transfusions simply delay episodes of splenic sequestration to a later age

Long-term management of splenic sequestration in children with sickle cell disease

J Pediatr 1990;117:194-9

Thomas R. Kinney, MD, Russell E. Ware, MD, William H. Schultz, PA-C, and Howard C. Filston, MD

From the Departments of Pediatrics and Surgery, Duke University Medical Center, Durham, North Carolina

Observed arm:
- 4/7 had recurrence
- 3/4 had splenectomy

Table II. Group 3: transfusion program patients (all with sickle cell anemia)

Patient No.	Recurrence		Duration of TX (mo)	Time to recurrence (mo)	Hemoglobin S at recurrence (%)	Therapy for recurrence	Time off TX (yr)	Duration of follow-up (yr)
	On TX	Off TX						
8	Yes	NA	10	9	16	S	1.7	2.6
9	Yes	NA	6	5	12	S	0.7	1.2
10*	Yes	NA	12	3	NR	S	3.1	4.0
11	No	Yes	12	1	35	S	5.1	6.0
12	No	Yes	9	3	NR	S	1.8	2.5
13	No	Yes	7	2	NR	S	5.8	6.4
14	No	Yes	6	2	43	S	1.5	2.0
15	No	No	6	NA	NA	NA	1.2	1.7
16	No	No	9	NA	NA	NA	4.0	4.7
17	No	No	6	NA	NA	NA	1.9	2.4
18	No	NA	Ongoing	NA	NA	NA	NA	0.8
19	No	NA	Ongoing	NA	NA	NA	NA	0.6
4†	Yes	NA	10	3	31	S	7.3	8.8
5†	No	Yes	6	1	NR	S	7.3	11.2

TX, transfusion; *NA*, not applicable; *S*, splenectomy; *NR*, not recorded.
*Noncompliant.
†Transfusion program begun after initial period of observation.

Transfused arm:
- 4/14 had recurrence ON Tx
- 5/8 had recurrence OFF Tx

Thus suggest that while splenic sequestrations can occur on chronic tx, rate is much lower but 61% of patients still required splenectomy

Splenectomy

- Recommended after first acute splenic sequestration episode or a minor episode if hypersplenism is sustained
- Splenectomy before age 2yrs is higher risk, but overall found no significant increased risk of infection with proper vaccination and antibiotic prophylaxis

Postsplenectomy course in homozygous sickle cell disease

J. G. Wright, MB, MRCP, MRCPath, I. R. Hambleton, BA, MSc, P. W. Thomas, PhD, N. D. Duncan, DM, S. Venugopal, MS, FACS, FICS, and G. R. Serjeant, MD, FRCP

J Pediatr 1999;134:304-9

130 patients (1974-1996)

35% after 2 splenic sequestration attacks
65% hypersplenism

Table I. Deaths in observation period in splenectomized group and control group (splenectomy group ranked by age at splenectomy, control group by age at death)

	Splenectomy			
Patient No.	Reason	Age	Age at death	Cause of death
Splenectomy group				
1S	ASS	1.3	6.8	Aplastic crisis
2S	ASS	2.9	10.1	Cerebrovascular accident
3S	ASS	3.1	3.8	Acute chest syndrome
4S	CHS	4.6	7.1	Unknown (PM not performed)
5S	CHS	4.9	5.0	No information available
6S	CHS	5.4	13.8	Acute chest syndrome
7S	CHS	10.9	11.3	Gastroenteritis, pulmonary edema
8S	CHS	10.9	11.2	Fever, acute hepatic sequestration
9S	CHS	13.8	27.4	*Salmonella enteritidis* septicemia

Splenectomy at mean of 2.3 years

Mortality was not increased in splenectomy group (9/130) compared to control group (12/130)

Table II. Bacteremia in splenectomy and control groups (splenectomy group ranked by age at splenectomy, control group by age at bacteremia)

	Splenectomy			
Patient No.	Reason	Age	Age at bacteremia	Organism
Splenectomy group				
1S	CHS	1.1	1.6	*Streptococcus pneumoniae*
2S	CHS	1.4	7.5	*Streptococcus pneumoniae*
3S	ASS	2.1	13.1	*Clostridium welchii*
4S	ASS	2.5	5.0	*Streptococcus pneumoniae*
5S	ASS	2.8	6.9	*Enterobacter*
6S	ASS	3.9	7.4	*Salmonella enteritidis*
7S	ASS	4.4	5.4	*Haemophilus influenzae* type b
8S (9S in Table I)	CHS	13.8	27.4	*Salmonella enteritidis* (died)
9S	CHS	20.8	27.6	*Streptococcus pneumoniae*
10S	ASS	25.4	29.7	*Escherichia coli*

Bacteremia was not increased in splenectomy group compared to control group

BUT increased Strep bacteremia vs Salmonella bacteremia in splenectomy group

Outcome of splenectomy in children younger than 4 years with sickle cell disease

J of Pediatric Surg (2009) 44, 1134-1138

Aaron P. Lesher[a], Ram Kalpatthi[b], Joshua B. Glenn[a], Sherron M. Jackson[b], Andre Hebra[a,*]

[a]*Surgery, Medical University of South Carolina, Charleston, SC 29425, USA*
[b]*Pediatrics, Medical University of South Carolina, Charleston, SC 29425, USA*

53 patients <4yrs old underwent splenectomy after 1 or more acute splenic sequestration episodes (1993-2008)

Table 1 Operative details of children with SCD (aged <4 years) who underwent splenectomy

Parameter	Laparoscopic[a] (n = 31)	Open (n = 22)	P[b]
Age at operation (y)	2.2	2.1	.69
Male sex (%)	15 (48.3)	13 (59.9%)	.45
Mean spleen weight (SD) (g)	100.6 (46.9)	120 (54.5)	.18
Mean length of hospital stay (SD) (d)	3.6 (1.2)	3.8 (1.3)	.84

[a] One patient underwent open splenectomy after laparoscopy (see text).
[b] Changes in mean levels analyzed using 2-tailed Student's t test with significant $P \leq .05$.

- Mean follow-up was 5.6yrs
- 3/53 died and 1/53 (1.8%) had pneumococcal sepsis
- All patients had pre-splenectomy vaccination and post-splenectomy PCN
- All patients admitted for 48hr R/O if febrile until 12yrs of age

- Conclusion: splenectomy at young age is safe and without increased risk