A Case of Overwhelming Post-Splenectomy Sepsis in a Vulnerable Child

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Introduction:

- Overwhelming Post-Splenectomy Infection (OPSI) is a potentially fatal complication following splenectomy
- *Streptococcus pneumoniae* is the leading causative organism, accounting for up to 50% of these cases[1]
- Discrepancies exist between regional recommendations for vaccination and other strategies to ameliorate this risk
- Asplenics account for <1% of cases of invasive pneumococcal disease in New Zealand, yet improved vaccine and antibiotic prophylaxis use could reduce this further

Case:

History

- 8 year old Samoan boy with hereditary spherocytosis and splenectomy aged 6yrs
- 6 hour history of fever with headache, rigors, vomiting and diarrhoea
- No regular medications or allergies
- Fully immunised per standard schedule. Partially immunised for asplenia protection, with polysaccharide pneumococcal (PPV23) and meningococcal vaccine but no conjugate pneumococcal vaccine (PCV)
- Family history of hereditary spherocytosis and splenectomy in Mother

Presentation

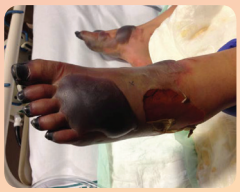
- Fever, poor perfusion, HR 99/min, tachypnoea and hypotension (initial BP: 81/23)
- On examination, depressed GCS and impetigo on legs
- Elevated creatinine (148), coagulopathy (INR 1.6) and metabolic acidosis (lactate 8)

Management

- Aggressive fluid resuscitation (60ml/kg crystalloid) and IV antibiotics
- Intubation and multiple inotropic supports for progressive shock
- *Streptococcus pneumoniae* on blood culture after 6 hours incubation
- Antibiotics initially ceftriaxone and flucloxacillin; changed to IV vancomycin, clindamycin and penicillin on D3. Later rationalized and completed 2/52 IV penicillin

Progress and Sequelae

- Purpura fulminans of hands and feet (image below)
- Ventilatory support until D15 of illness
- DIC from onset, needing multiple blood product support
- Dialysis (CVVH) on D3-14 for acute kidney injury



- Chest drain for effusion on D4 , then thoracotomy for a complication of insertion
- Left leg cellulitis on D12
- Line sepsis D16
- Upper GI bleeding D16-21 from jejunal ulceration.

Outcome:

- Good recovery
- Dry necrotic toes, requiring plastic surgical input
- Plan for lifelong penicillin prophylaxis, standby antibiotics and further immunisations (PCV13, Hib, MCV-4D)
- *S. pneumoniae* further serotyped as 23B

Discussion:

- Strategies for reducing sepsis risk post-splenectomy include patient education, antibiotic prophylaxis, early administration of stand-by antibiotics and immunisation for encapsulated organisms (*S. pneumonia, Haemophilus influenzae* type *b*, meningococcus) and influenza
- Recommendations for pneumococcal immunisation differ in timing of doses and use of conjugate pneumococcal vaccine (PCV) in adults (Table 1)
- Vaccination with PCV prior to PPV23 results in higher antibody responses for their common serotypes. The evolution of PCVs and changing availability/ eligibility criteria in NZ (Figure 1) has resulted in some asplenics, including our patient, being suboptimally protected
- More than half of asplenics with invasive pneumococcal disease in New Zealand had not received any pneumococcal vaccinations (Table 2)[2].
- Our case was due to *S.pneumoniae* serotype 23B, which is not present in any available vaccine. Crossprotection within some vaccine serotypes (eg for 6A by 6B) has been demonstrated4, though not specifically in 23B
- Guidelines differ regarding the duration of antibiotic prophylaxis, and whether it should be lifelong for all risk groups (Table 3)
- Our patient had discontinued penicillin prophylaxis. Although he was outside the 2 year post-splenectomy period, longer prophylaxis may have been appropriate as he had an underlying haematological condition

TABLE 1: Pneumococcal Vaccination Recommendations in Asplenia/Splenectomy

	Australia (2014)[3]	NZ (2014)[4]	BCSH (2011)[5]	IDSA (2013)[6]
Normal Schedule	PCV10/13 at 2, 4 and 6mo. PCV13 in primary course for high risk. Booster at 12-15mo if PCV10 or high risk.	PCV13 at 6wk, 3, 5 and 15mo.	PCV13 at 2, 4 and and 13mo.	PCV13 at 2, 4, 6 and 12-15mo (as per CDC).
<1yo	As above	As above. PCV13 booster if primary course with PCV7/10.	As above.	As above.
2-5yo	1 dose of PCV13 if no booster after primary course. 2 doses of PCV13 if primary course not completed.	2 doses PCV13, 8 weeks apart for previously unimmunised. PCV13 booster if primary course with PCV7/10.	PCV13 booster if primary course with PCV7. 2 doses of PCV13 if partially or unimmunised.	1 dose of PCV13 if completed primary course PCV (3 doses by 2yo). 2 doses of PCV13 otherwise.
5-18yo	1 dose of PCV13 if nil previous.	1 dose PCV13.		1 dose of PCV13 if nil previous.
>18yo	1 dose of PCV13 if nil previous.	1 dose of PCV13 (not funded).		1 dose of PCV13 if nil previous.
All	Give dose of 23PPV when >4-5yo, and repeat 5y later.	Give dose of 23PPV when >2yo, and repeat 5y later.	Give dose of 23PPV when >2yo	Give dose of 23PPV when >2yo, and repeat 5y later.

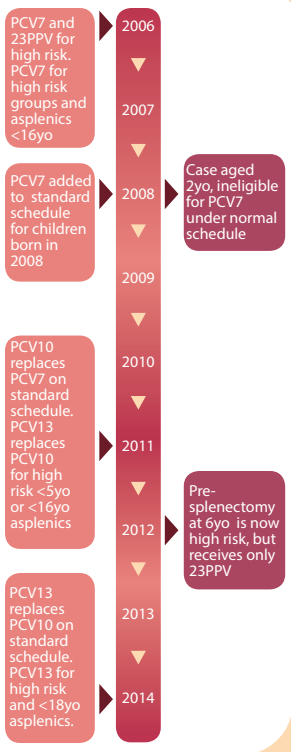
TABLE 2: Cases of Invasive Pneumococcal Disease in New Zealand

	2009	2010	2011	2012	2013
Total	647	535	552	488	479
Number asplenic	6	4	5	1	7
Where vaccine status known, # asplenics with no hx of any pneumococcal vaccine	2	3	3	1	3

TABLE 3: Antibiotic Prophylaxis Recommendations for Asplenic Patients

	Year	Patients at Increased Risk ("High Risk")	Antibiotic Prophylaxis Recommended	Patient Initiated Treatment of proven/suspected Infection
ASID[1]	2008	• <2y post-splenectomy • Immune suppression.	For 2y post splenectomy. Lifelong antibiotics for high risk. Lifelong antibiotics for low risk, if patient accepting.	HD amoxycillin, augmentin, cefuroxime or moxifloxacin (if pencillin allergic).
BCSH[5]	2011	• <16yo and >50yo • Poor pneumococcal vaccine response • Hx invasive pneumococcal disease (IPD) • Immediately after splenectomy for trauma • Haematological malignancy	Lifelong for all high risk patients, and consider same for low risk patients.	According to local protocols.
Canadian Paediatric Society[7]	2014	• <5yo • <2y post-splenectomy • Hx IPD	For 2y post splenectomy. For all high risk. Ideally, for low risk patients also.	Seek medical attention.
Starship[8]	2014	• Younger children • <2yr post-splenectomy • Haematological disorder • Immune suppression	Under discretion of ID/ haematology.	HD oral amoxycillin (90mg/kg/day) and seek medical attention.

FIGURE 1



Conclusion:

Invasive pneumococcal disease is a life long risk post-splenectomy, and does occur outside the traditional high risk cohort. Patient education, early medical attention, appropriate antibiotic prophylaxis and completion of recommended immunisations is important. The difficulties of keeping clinicians and patients informed of changing recommendations could be addressed by the development of a New Zealand asplenia registry.

References:
1) Spelman, D., J. Buttery, A. Daley, D. Isaacs, I. Jennens, A. Kakakios, R. Lawrence et al. "Guidelines for the prevention of sepsis in asplenic and hyposplenic patients." *Internal medicine journal* 38, no. 5 (2008): 349-356.
2) "Invasive Pneumococcal Disease Reports." Public Health Surveillance. December 30, 2015. https://surv.esr.cri.nz/surveillance/IPD.php.
3) *The Australian Immunization Handbook 2013*. Canberra: Australian Department of Health.
4) Immunisation Handbook 2014. Wellington: Ministry of Health, 2014.
5) Davies, John M., Michael PN Lewis, Jennie Wimperis, Imran Rafi, Shamez Ladhani, and Paula HB Bolton-Maggs. "Review of guidelines for the prevention and treatment of infection in patients with an absent or dysfunctional spleen: Prepared on behalf of the British Committee for Standards in Haematology by a Working Party of the Haemato-Oncology Task Force." *British journal of haematology* 155, no. 3 (2011): 308-317.
6) Rubin, Lorry G., Myron J. Levin, Per Ljungman, E. Graham Davies, Robin Avery, Marcie Tomblyn, Athos Bousvaros et al. "2013 IDSA clinical practice guideline for vaccination of the immunocompromised host." *Clinical infectious diseases* (2013): cit684.
7) Salvador, Marina L, and Victoria E. Price. "Preventing and treating infections in children with asplenia or hyposplenia." *Paediatrics & child health* 19, no. 5 (2014): 271-278.
8) Wilson, Elizabeth, Rachel Webb, Nyree Cole, Jane Skeen, James Hamill, Lochie Teague, Helen Evans, and Tim Prestige. "Splenectomy." Starship Guidelines: Splenectomy. July 4, 2014. Accessed December 30, 2014.
9) Park, In H., Matthew R. Moore, John J. Treanor, Stephen I. Pelton, Tamara Pilishvili, Bernard Beall, Mark A. Shelly, Barbara E. Mahon, and Moon H. Nahm. "Differential effects of pneumococcal vaccines against serotypes 6A and 6C." Journal of Infectious Diseases 198, no. 12 (2008): 1818-1822.