

Prevention of infection in sickle cell
disease -
a clinician's approach

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The problem

- Increased risk from
 - Bacterial: *esp children .. but also adults*
 - strep pneumoniae : 400 –fold increase sepsis and 600 x risk of *invasive disease*
 - even after immunisation programme – 15 cases IPD / 10,000 / yr
 - H influenzae, ~ 4 X risk
 - Neisseria meningitidis
 - Salmonella, other gram-negative bacteria*
 - Staph aureus*
 - Viral
 - Influenza
 - Hepatitis B
 - Malaria

because

- Functional asplenia
- Reduced complement activity
- Failure to make specific IgG antibodies to polysaccharide antigens

S. pneumoniae, Hib, N. meningitidis, E. Coli, Salmonella, Klebsiella, Group B streptococci



Risk has driven the screening programmes.

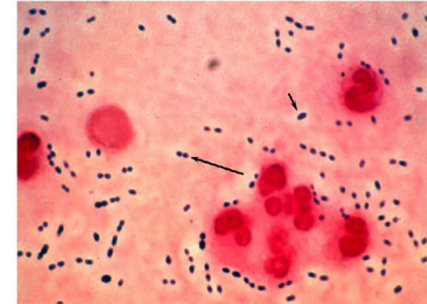
Risks greater for HbSS and HbS β 0 .. BUT same usually applied to all.

SO?

- Antibiotic prophylaxis
 - Immunisation
- Hydroxycarbamide?
- Early presentation

➤ Antibiotic prophylaxis

- ‘the single most important study in SCD in 20 years’
 - Penicillin bd prevents 84 % life-threatening episodes of strep pneumoniae in children [Gaston et al, N Eng J Med 1986]
 - Penicillin bd from newborn [US]
 - 90% by 3 months, 99% by 6 months [UK]
 - Doses differ! [US 125 mg bd 0 – 3 then 250 mg bd]
 - Erythromycin for penicillin allergic
- **But when to stop?**



- Initial study – safe to stop at age 5
- Follow up study
 - continuing vs placebo
 - 3 years follow up
 - No difference in pneumococcal or meningococcal infections [v small in both]
- Increased resistance ?
 - Some studies + / some –
 - US: can stop at 5 years, ***as long as***
 - Fully vaccinated, no previous IPD, not splenectomised
 - UK: usually continue into teens – or beyond if fully adherent
 - Check at least annually
 - If taking ‘some days’ - STOP

Antibiotics 'on hand'

- No actual evidence, but concensus..
 - carry at all times a treatment dose to start 'at first hint of infection, sore throat, fever'
 - Penicillin V 500 mg qds
 - Amoxycillin 500 mg tds
 - Not co-amoxyclav – often 1st line if admitted
- If not better within 24 hours – present for assessment / consideration broader spectrum antibiotics
- If temperature > 38 Ccome to hospital!

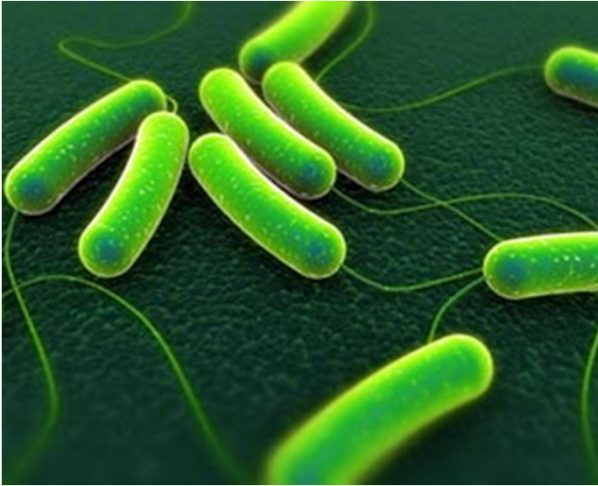
BUT .. continue regular bd dosing

IF

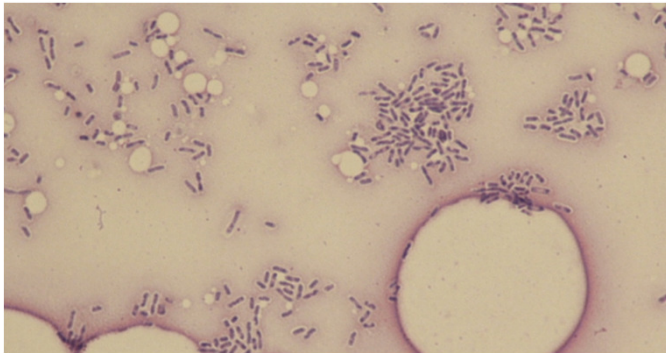
- Splenectomised
- Previous invasive pneumococcal infection

- And probably after age 50

Another one to prevent...



- Food hygiene
- Low threshold for suspicion
- Culture!
- Treat – always
- Rapidly progressive multi-organ failure, DIC, mortality high.



Food safety for people with sickle cell disease.

DIETARY ADVICE FOR PEOPLE WITH SICKLE CELL.

Having Sickle Cell disease affects your ability to fight infections. For this reason it is important for you to take certain precautions to reduce your risk of a number of infections. You will have been given a supply of antibiotics, and have had extra vaccinations, to help prevent some important infections. **HOWEVER**, these do not cover all infections and we want you to be aware, too, of others which you can get from certain foods. Being careful about what you eat could help protect you against some potentially dangerous infections. One of these, *salmonella*, can be caught from eating undercooked eggs or chicken.

FOODS YOU SHOULD AVOID

- ◆ Uncooked or undercooked ready - prepared meals. Make sure you heat ready-prepared meals until they are **pipng hot** all the way through.
- ◆ **Chicken** and other poultry should be thoroughly cooked, with no sign of remaining 'pinkness' in the meat.
- ◆ Raw eggs or food **containing raw or partially cooked eggs** (Tiramisu, Mayonnaise). Only eat eggs that are cooked until the white and the yolk are solid.

SOME OTHER GOOD FOOD HYGIENE AND GENERAL ADVICE which will help to reduce your risk of infections

- ◆ Always wash your hands after handling raw meat and poultry (chicken).
- ◆ Make sure you only eat meat that has been well cooked. Take particular care with sausages and mince meat.
- ◆ It is very important to keep certain foods at the right temperature to prevent bacteria growing or toxins forming. Always look at the label, and if it says that the food needs to be refrigerated, make sure you keep in the fridge.
- ◆ Cooked leftovers should be cooled quickly and put in the fridge.
- ◆ Keep raw and ready to eat foods separate
- ◆ Store raw meat in sealable containers at the bottom of the fridge, so it can't drip onto other foods
- ◆ Use different chopping boards/work surfaces for raw food and ready-to-eat foods
- ◆ Clean knives and other utensils thoroughly after use with raw food

AND ALSO...

- ◆ Always wear gloves when you are gardening or changing cat litter, and wash your hands afterwards.
- ◆ Person-to-person spread is reduced by thorough hand washing after going to the toilet or handling soiled clothes eg: changing babies' nappies
- ◆ Don't handle food when you are ill with stomach problems, such as diarrhoea or vomiting
- ◆ Don't handle food if you have sores or cuts, unless they are covered with a waterproof dressing
- ◆ Avoid keeping reptiles eg: lizards and snakes as pets as many of these have Salmonella and can spread infection.

Acknowledgement:
Food Standards Agency

◆ **Be safe!**
Reduce your risk of infection from food.

➤ Immunisations.

Pneumococcal conjugate vaccines
PCV:

- Prevnar 7 – 2000
- Prevnar 13 – 2010
- Those now aged over 18 won't have received:
 - **GIVE**
- Any who have only had Prevnar 7 ... **give** 1 dose Prevnar 13

- *Note . . . Case numbers non-vaccine serotypes are increasing.*

- Pneumococcal polysaccharide vaccination PPV23 'Pneumovax'
 - At five yearly intervals
 - Or just 1 boost ?
 - *Repeated dosing might be counterproductive . . . But remains recommendation in UK*
 - Gap needed between the two
 - Pvax 8 weeks after PCV
 - PCV 6 months after Pvax

Others - bacterial

- If not immunised according to UK schedule as infants.. also
 - Hib / Men C – 1 dose
 - MenACWY – 1 dose, 1 month later
 - MenB – two doses, 1 month apart [can be with others]
 - ? Include recommendation as a 'PS' in clinic letters?

Immunisation against viruses

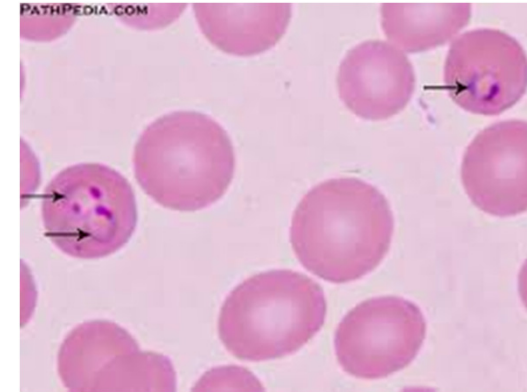
- Many – eg parvovirus ... can't
- Can – and recommended:
 - 'flu
 - Hepatitis B series
 - Risk from transfusion, low – but avoidable
 - Check antibody titres and boost as needed.

Where to give?

- Mostly primary care
 - **EXTREME DIFFICULTY** getting confirmation!
 - In clinic:



malaria



HbAS – reduced parasite density and less severe malaria BUT mortality rate in HbSS is HIGHER than others

Remember! advise prophylaxis and early treatment . .

[and all recommended additional including live vaccines / yellow fever etc]

Take course of co-amoxyclav / ciprofloxacin if area services sparse.

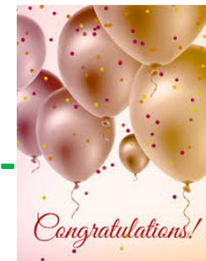
➤ Hydroxycarbamide

- Regard as ‘adjunct to reducing infection in SCD’.
 - High levels of HbF in childhood may help preserve splenic function / reduce susceptibility to encapsulated bacteria.
 - Sickle mice on HC : reduced progression of IPD
 - Down regulation E-selectin [Lebensburger, Blood, 2012]
 - Reduced leucocytosis and inflammation.
- Offer to all HbSS / Hb S β_0 thalassaemia
- Recommend to any ‘ ‘ with overt complications.

➤ Early presentation



- Check / remind at least at annual review
- Present if 38 C or above [some guidelines say 38.5 ..]
- **Say this until you are tired of hearing yourself saying it!**
- **As well as** ‘so if you get a sore throat, fever, feel shivery, cough, other signs of infection – what are you going to do’?
 - **Take my temperature**
 - **Start on my antibiotics if temperature below 38 C**
 - **Come to hospital if temperature above 38 C ... YES! - - -**



Thank you.