Asplenia

Presented by :

Eman Ahmad Al-Omoush

The role of the spleen in the immunity

- Spleen has mainly two function :

1) **Lymphoid** function (carried out by the white pulp).

2) **Filtration** of blood (by the red pulp).

The case of Susan Vaderveer

- **Parents were distant relatives**
- **10 months old**
- **She developed a cold for 2 weeks**
  - she became sleepy (fatigued)
  - felt very hot (high temperature)
  - she started having convulsions
- **Died on the way to the hospital**
- **Postmortem cultures grew haemophilus influenza, type b**
- **Autopsy revealed : she had no spleen**

Siblings

- **Betsy (3 yr old)**

-she had fever, complained of an earache and her eardrums were found to be red.

-her WBC count was very elevated. (28.500cells/microliter)

-cultures grew out Haemophilus influenza, type b.

-she was given IV ampicillin for 10 days in the hospital.

-In the following year she was seen by a pediatrician 3 times for otitis media, pneumonia and mastoditis.

- **David (5 yr old)**

  -at 21 months of age he had meningitis caused by Streptococcus pneumoniae.

  -at 27 months of age he had another occurrence of pneumococcal meningitis.

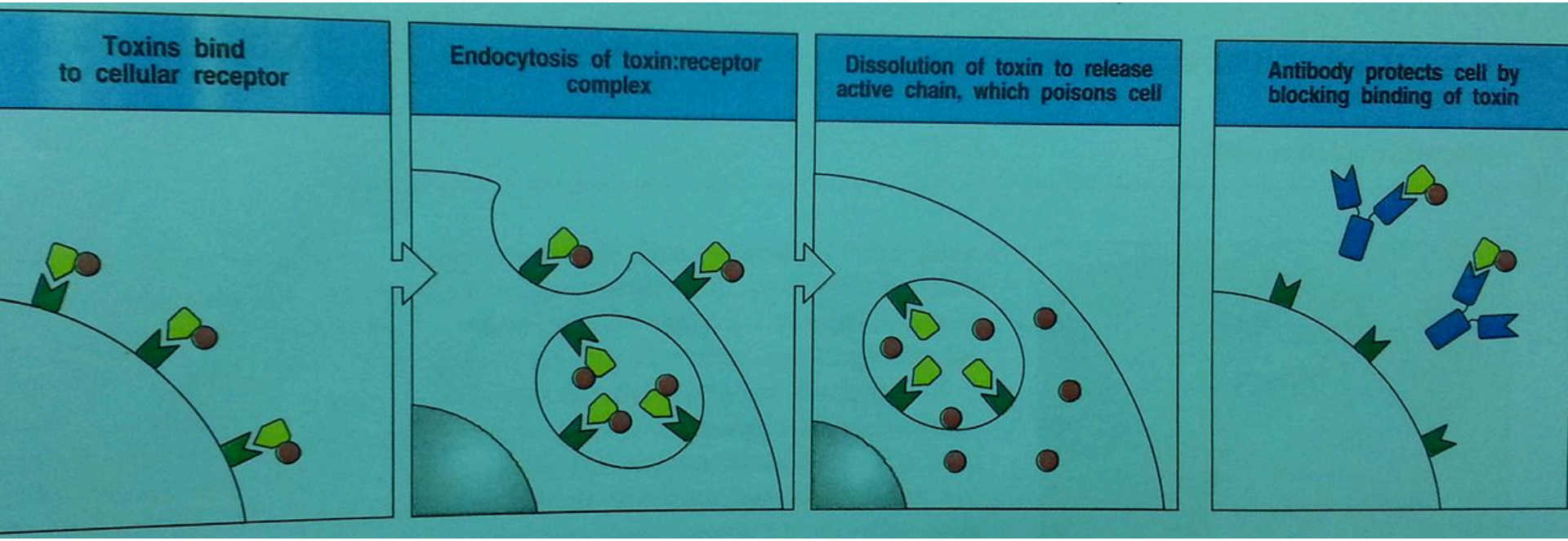
  -at age of 3 years he had had pneumonia.

  -at the time of Susan's death he was well

The two other children of the Vandeveer, a girl aged 8 years and a newborn male, were in good health.

- All Vaderveer children were immunized against tetanus, diphtheria and whooping cough at ages 3, 4 and 5 months.

- Agglutination tests to diphteria, tetanus and pertussis toxins were normal.

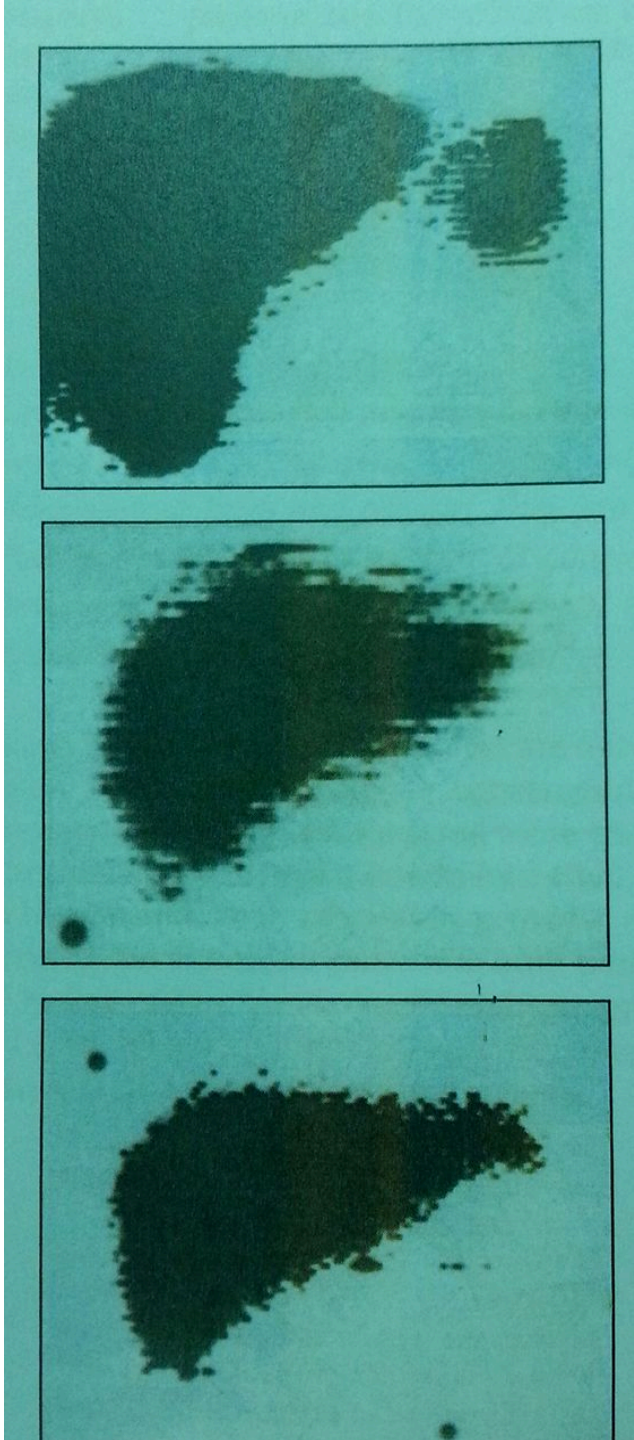


- Neutralization by antibodies protects cells from toxin action.
- In the case of the DPT vaccine, the killed bortedella pertussis cells act as an adjuvant, which enhances the immune response to all of the components of the vaccine by delivering activating signals to antigen-presenting cells.

Siblings	age	Tetanous Toxiod	Typhoid vaccine subcutaneously	1ml 25% SRBC IV	2-4 weeks after SRBC	Radioactive test results
Betsy	3	1:32	1:32	1:32	No Increase	No Spleen present
David	5	1:32	1:16	1:4	No Increase	No Spleen present
Girl	8	Similar	1:32	1:32	1:256	Spleen is present

•All the children and their parents were injected intravenously with radioactive colloidal gold, which is taken up by the reticuloendothelial cells of the liver and spleen within 15 minutes after the injection. A scintillation reveals that Betsy and David have no spleens !

*Why did David and Betsy have normal responses to the typhoid vaccine but not to the sheep red blood cells ?
Typhoid vaccine was given subcutaneously where regional lymph nodes produces an immune response. But SRBCs were given IV and in the case of Betsy and David we found that they don't have spleens to fight these foreign cells thus there won't be an immune



- The genetic defect causing asplenia has not been identified.

- The Vaderveer family is unusual in that 3 of their first 4 children were born without spleens. After the events described in the case, they had 3 more children. One of the boys and the girl were also born without spleens.

- The major consequence of its absence is a susceptibility to bacteremia, usually caused by the encapsulated bacteria Streptococcus pneumonia or Haemophilus influenza.

- This susceptibility is caused by a failure of the immune response to these common extracellular bacteria when they enter the bloodstream.

- Congenital asplenia is not very common but splenectomy due to rupture of the spleen or malignancy in the spleen is more common.

- After splenectomy, patients, particularly children are susceptible to bloodstream infections by microorganisms to which they have no antibodies.

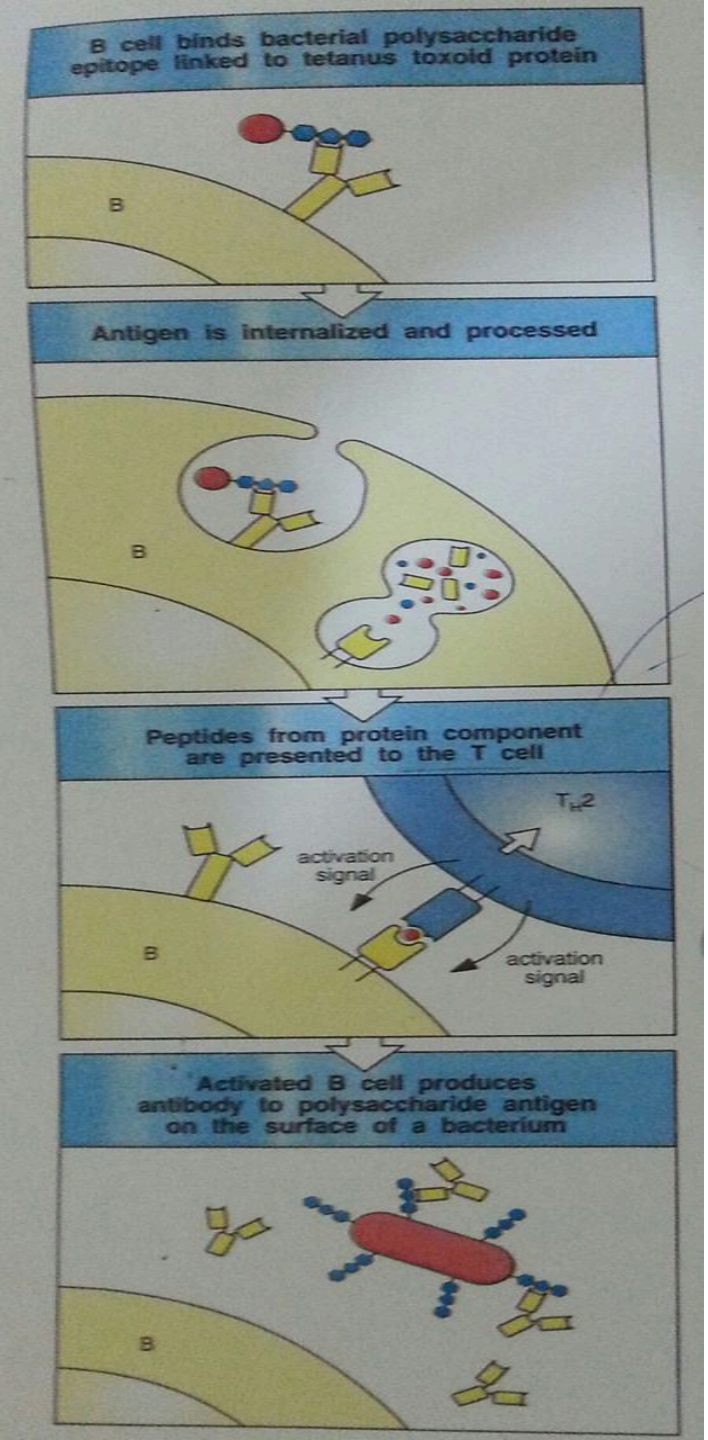
- Adults who already have antibodies to these microorganisms are therefore much less vulnerable to problems of bacteremia than children who have not yet developed antibodies to these germs.

- **Immunology consultation ??**

  -Hib vaccine

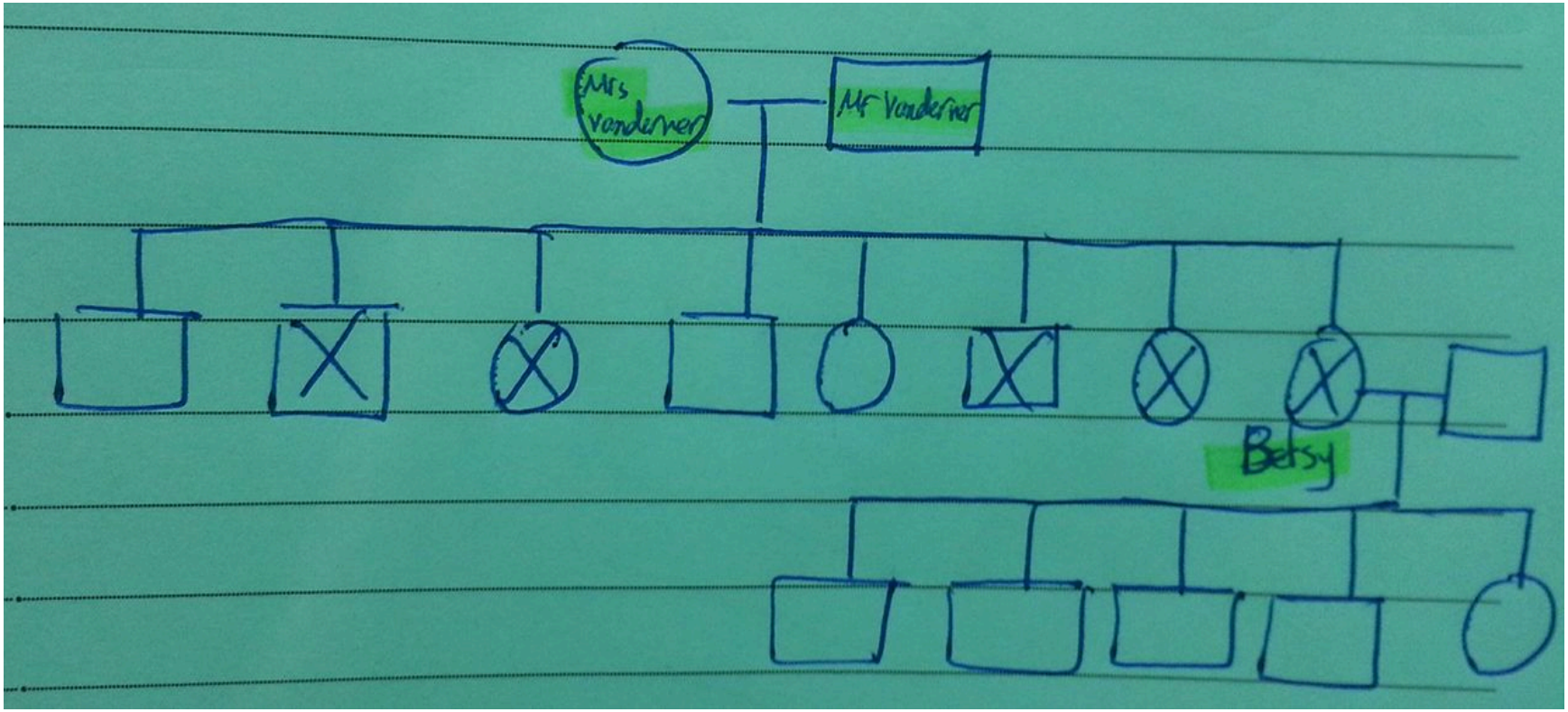
  -Pneumovax (a vaccine containing the major prevalent pneumococcal polysaccarides)

  -prophylactic antibiotics (taken at low doses daily but at higher when there is any dental work)



•Haemophilus influenza b vaccine is a conjugate of bacterial polysaccharide with tetanus toxoid protein, which enhances the immune response by allowing a polysaccharide-specific B cell to recruit T-cell help.

The B cell recognizes and binds to the polysaccharide, internalizes and degrades the toxoid protein to which it is attached, and displays peptides derived from it on surface of MHC class II molecules. Helper T cells generated in response to earlier vaccination against the toxoid recognize the complex on the B cell to produce antibody against the polysaccharide. This antibody can then protect against infection with H.Influenza type b.



Congenital Asplenia is an autosomal recessive disease.
(it affects both sexes and there is skipped generation)

Thank you !