



# A case of two siblings with fatal invasive pneumococcal disease

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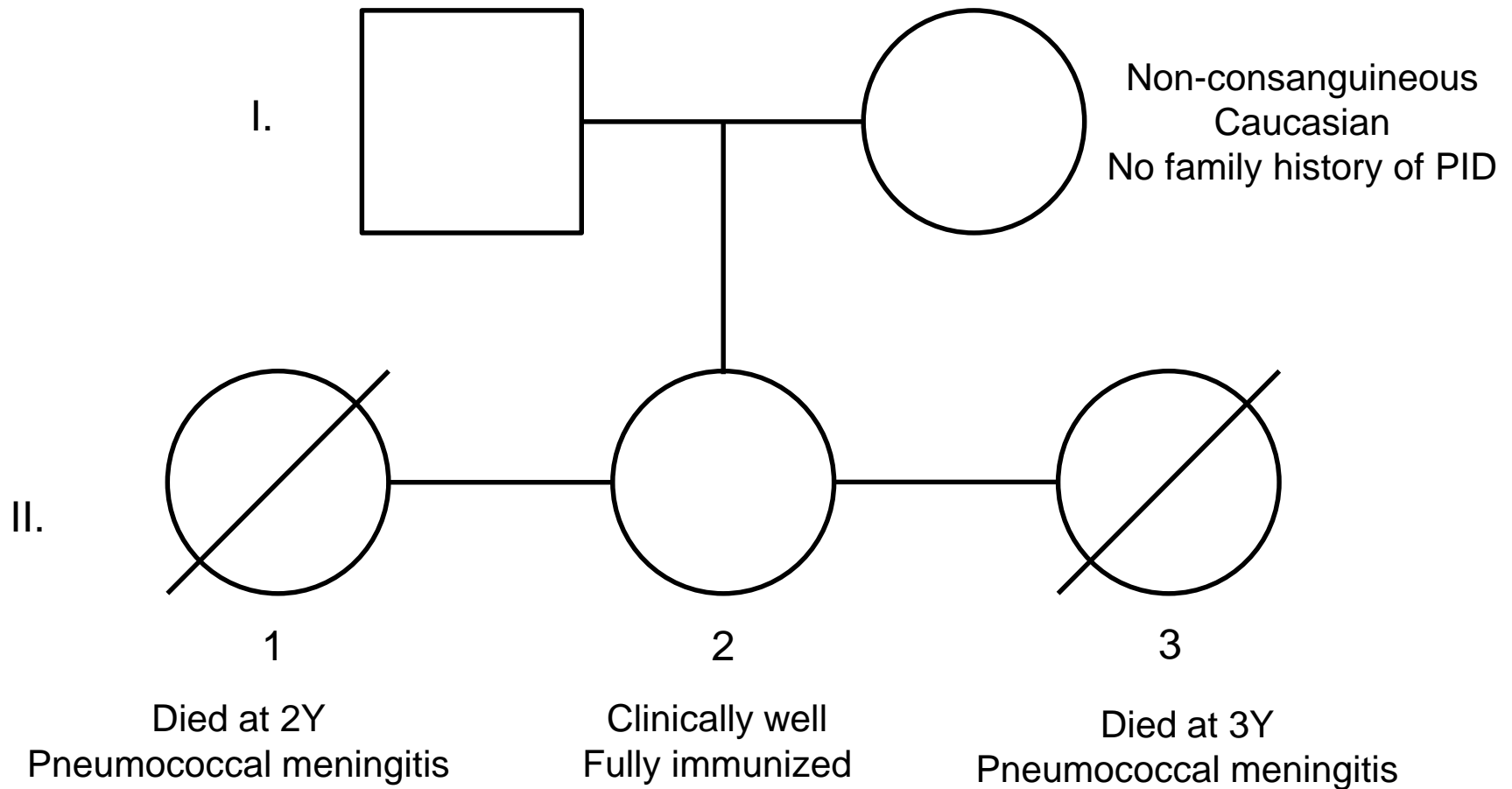
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# Overview of cases



# Clinical Presentation

## Patient 1

- 2YF with 3 days of fever
- Pneumococcal meningitis – Serotype 15C\*
- Arrest due to neurologic complications
- Past history of recurrent lymphadenitis
  - “Never very sick”
  - No fever
  - No erythema

## Patient 2

- 3YF with 24 hours of emesis, headache, lethargy
- Pneumococcal meningitis – Serotype 15A\*
- Brain death due to neurologic complications
- Unremarkable past history
  - Normal T/B subsets at birth
  - Immunized, no infections

*\*Not covered by PCV13*

*Consider the differential diagnosis:  
Which hypotheses and which tests?*



# Investigations

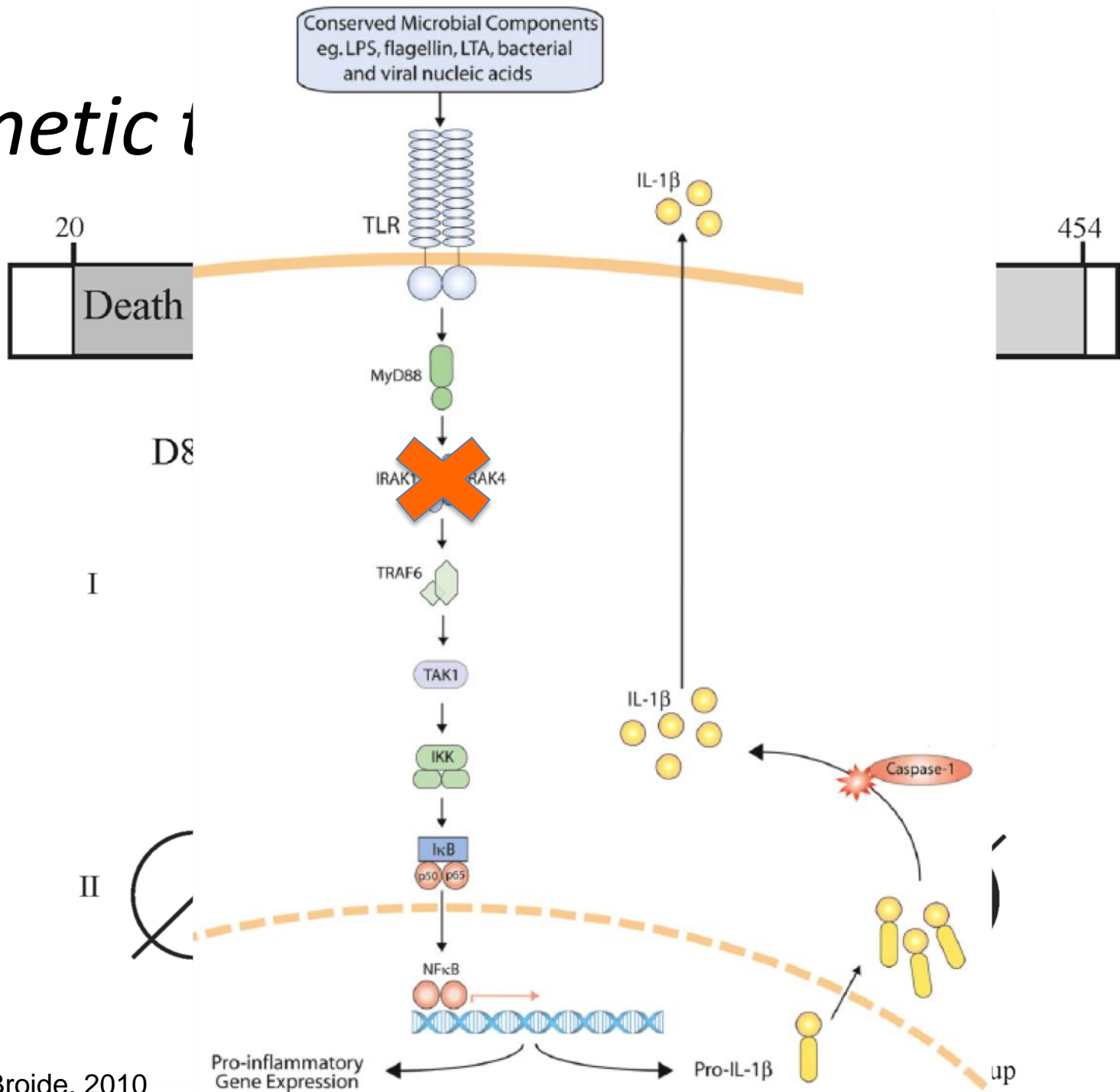
## Patient 1 (2YF)

WBC	6.8
Neutrophils	1.97
Lymphocytes	2.65
Smear	Poikilocytosis

## Patient 2 (3YF)

Done at 3Y	WBC	4.7 x 10 <sup>9</sup>	
	Neutrophils	4.15 x 10 <sup>9</sup>	
	Lymphocytes	0.47 x 10 <sup>9</sup>	
	Smear	Poikilocytosis	
	IgA	0.44 g/L	0.25-1.9
	IgG	4.73 g/L	4.5-13.5
	IgM	0.28 g/L	0.24-2.1
	CH50	30 CAE	63-145
	AH50	46x10 <sup>3</sup> /L	92-152
	Done at 4 months	CD3 cells	4.24x10 <sup>9</sup> /L
CD19 cells		1.06x10 <sup>9</sup> /L	0.6-3.0
CD3- CD56+ cells		0.31	

# Genetic t



## *Points for discussion*

- What clinical features are most suggestive of toll-like receptor signaling defects?
- In patients with a family history of fatal invasive pneumococcal disease, what immune workup should be done?



**a place of mind**

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