

Vasculitis of the young

Dr Paul Brogan

Academic Consultant in Paediatric Vasculitis
and Honorary Senior Lecturer

Great Ormond St Hospital and Institute of
Child Health, London UK

Overview

- Children are not little adults: case discussions
- Severe vasculitis in children is rare but does occur
- Specific issues relating to vasculitis and children/young people
 - Difficulties with diagnosis and classification
 - Difficulties with treatment
 - Difficulties with doing research
 - Effect on growth, schooling, rest of family
 - The future?

Case

Male, age 14

- 1st year of life: rash and cough
- Age 1: lung biopsy: patchy bronchiolitis with chronic alveolar and interstitial inflammatory infiltrate, no interstitial fibrosis; vasculitic lesions on ears-polychondritis on biopsy, and deep vessels demonstrating frank vasculitis

Relapsing polychondritis? (MJD)

Investigations of note

- Long-standing high ESR, CRP, and SAA Aug 2007 661 mg/L (0-9.9)
- Inconsistent positive: ACL (& DRVVT +); RF; cryoglobulins; (ANCA negative)
- Intermittent high TNF-alpha, TNFR1, and IL-1ra (peripheral blood); normal IL-6
- Intermittent high circulating endothelial cell counts

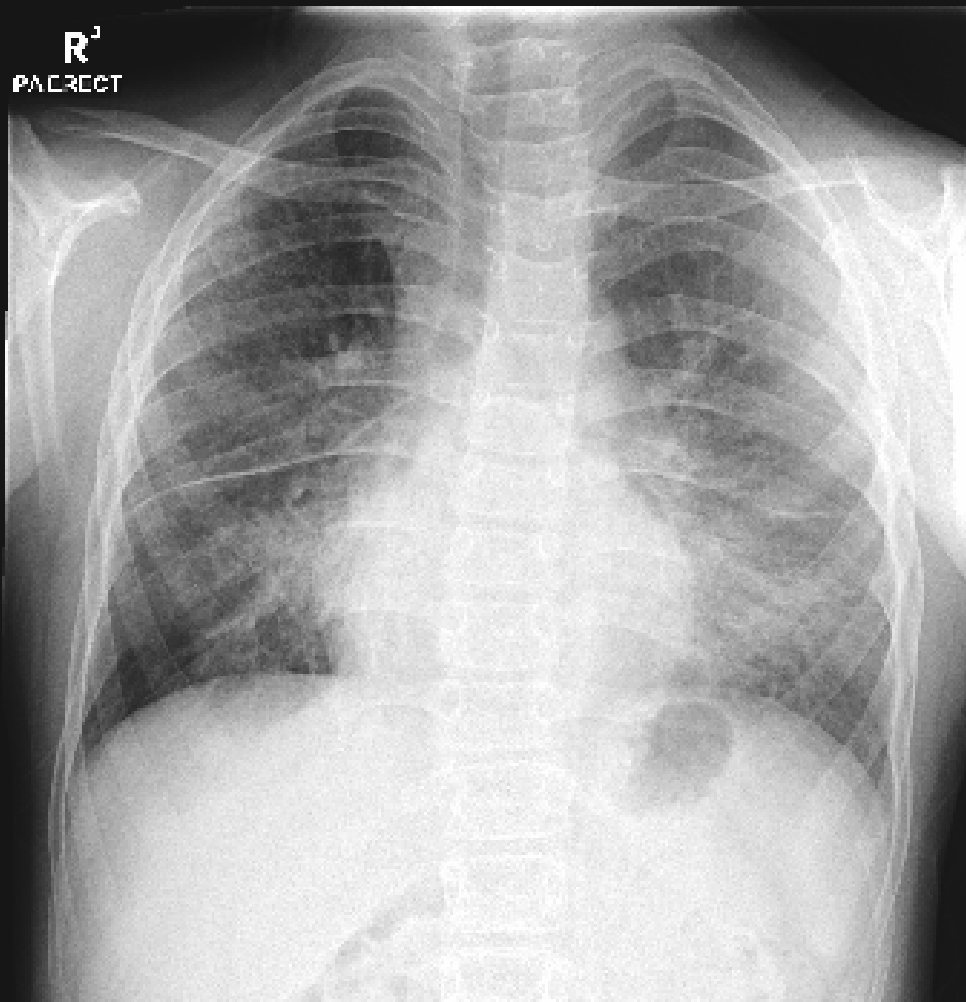
Therapy: age 2 years-14 years

- Age 2: corticosteroids
- Age 5: Cyclosporin- renal impairment (biopsy in 2002: glomerulosclerosis 5/17 glomeruli; no large vessels present, small vessels normal; no deposition of immunoreactants)
- Age 9: Oral Cyclophosphamide, then azathioprine

Therapy: age 2 years-14 years

- Feb 2005: Rituximab (750 mg/m²) X 2 with IV cyc 500mg
- No family match for Bone marrow transplant: Sep 2006
- October 2006- present: Infliximab 6mg/kg-6 weekly; prednisolone (0.2-0.5 mg/kg/day); azathioprine; nifedipine; sildenafil; growth hormone; sustinon

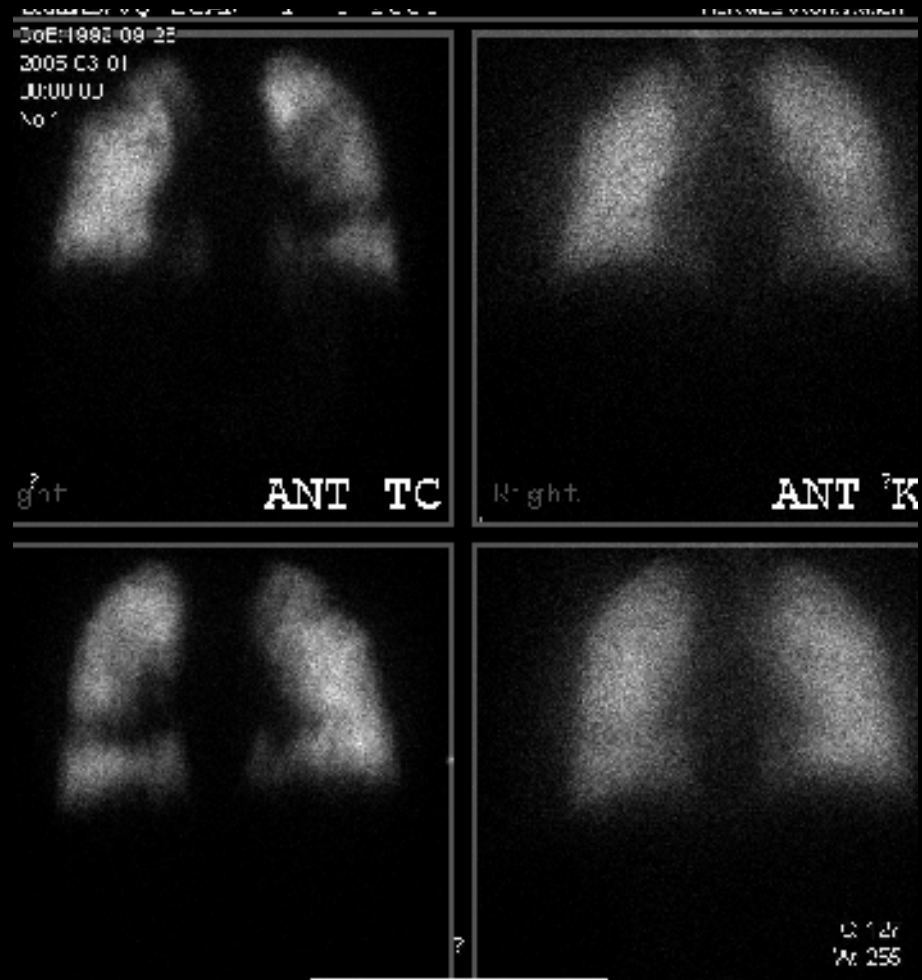
R¹
PA RECT

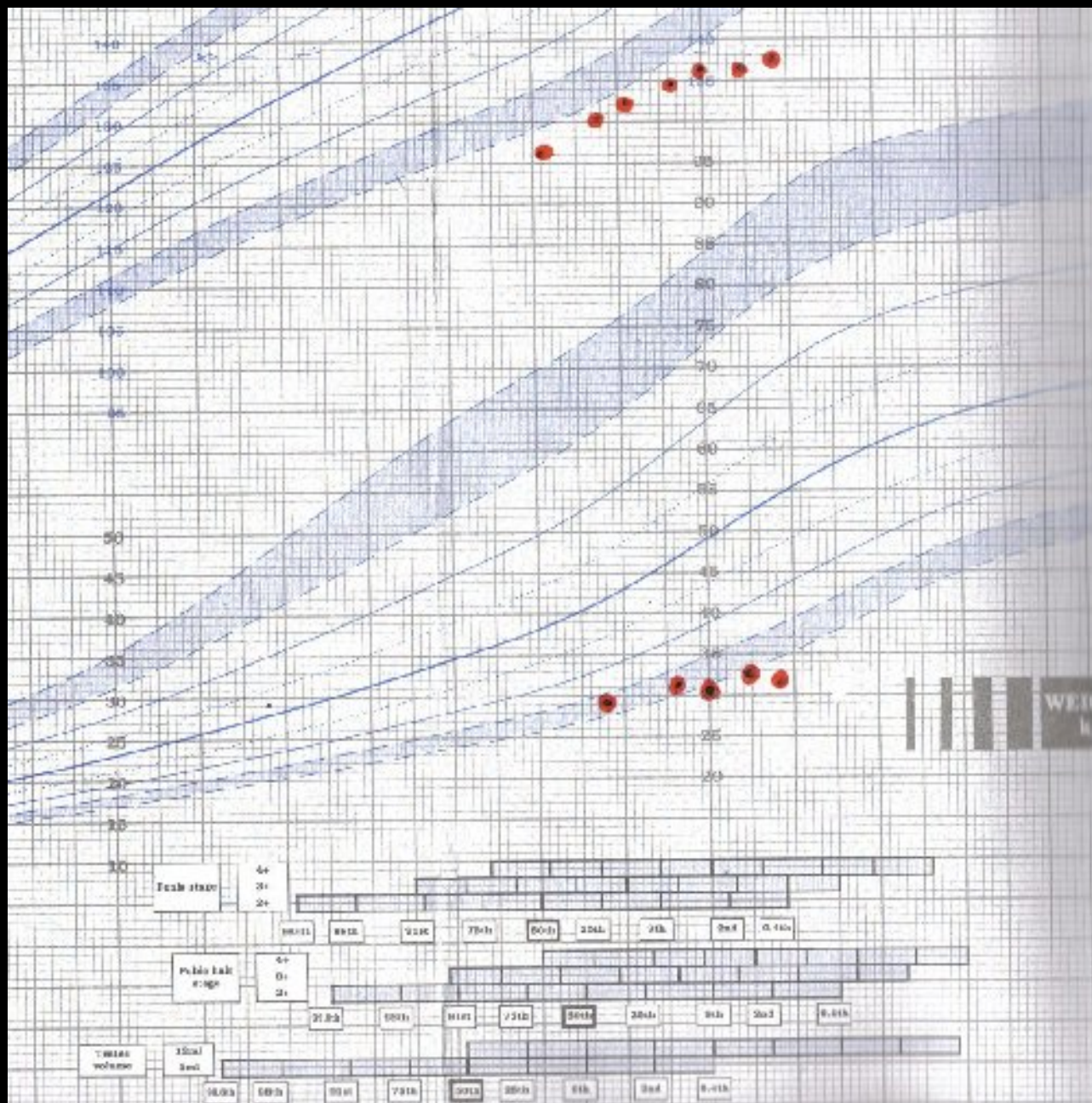


?

GREAT ORMOND STREET CH

V/Q scan





Currently

- Improved but still ongoing systemic inflammation (↑ amyloid)
- DEXA scan: Z score -3.5
- Moderate pulmonary hypertension
- FVC 44%; FEV1 36%
- Still B cell deplete 30 months post rituximab
- GFR:
- New onset arthropathy hands
- New onset palatal weakness ?mononeuritis

Discussion

- Diagnosis: RP, but small vessel vasculitis driving it? Doesn't fit into a neat category
- Growth
- School
- Effect on whole family
- Further treatment
- Transition to adult care