

## Recent peer-reviewed publications

### 2010

Vinh DC, Patel SY, Uzel G, Anderson VL, Freeman AF, Olivier KN, Spalding C, **Hughes S**, Pittaluga S, Raffeld M, Sorbara LR, Elloumi HZ, Kuhns DB, Turner ML, Cowen EW, Fink D, Long-Priel D, Hsu AP, Ding L, Paulson ML, Whitney AR, Sampaio EP, Frucht DM, DeLeo FR, Holland SM. Autosomal dominant and sporadic monocytopenia with susceptibility to mycobacteria, fungi, papillomaviruses, and myelodysplasia. *Blood*. 2010 115(8):1519-29.

### 2009

**Hughes SM**, Amadi B, Mwiya M, Nkamba H, Tomkins A, Goldblatt D. Dendritic cell anergy results from endotoxemia in severe malnutrition. *J Immunol*. 2009 183(4):2818-26

**Hughes SM**, Amadi B, Mwiya M, Nkamba H, Mulundu G, Tomkins A, Goldblatt D. CD4 counts decline despite nutritional recovery in HIV-infected Zambian children with severe malnutrition. *Pediatrics*. 2009 123(2):e347-51.

Soncini E, Slatter MA, Jones LB, **Hughes S**, Hodges S, Flood TJ, Barge D, Spickett GP, Jackson GH, Collin MP, Abinun M, Cant AJ, Gennery AR. Unrelated donor and HLA-identical sibling haematopoietic stem cell transplantation cure chronic granulomatous disease with good long-term outcome and growth. *Br J Haematol*. 2009 145(1):73-83.

Rodriguez-Marco A, Bradbury M, Riley P, **Arkwright PD**. Autoimmunity and recurrent infections in partial C3 deficiency. *Rheumatology* 2009 (accepted)

**Arkwright PD**, Luchetti F, Tour J, Roberts C, Ayub R, Morales AP, Rodríguez JJ, Gilmore A, Canonico B, Papa S Degli Esposti M. Fas stimulation of T lymphocytes promotes rapid intercellular exchange of death signals. *Cell Research* 2009 22 Sept [epub]

Hong M, Ryan K, **Arkwright PD**, Gennery AR, Costigan C, Dominguez M, Denning D, McConnell V, **Cant AJ**, Abinun M, Spickett GP, Swan D, Gillespie C, Young D, Lilic D. Pattern Recognition Receptor Expression is Not Impaired in Patients with Chronic Mucocutaneous Candidiasis with or without Autoimmune Polyendocrinopathy Candidiasis Ectodermal Dystrophy (APECED). *Clin Exp Immunol* 2009;156:40-51.

**Arkwright PD**. Omalizumab or rituximab for severe chronic autoimmune urticaria. *J Allergy Clin Immunol* 2009;123:510-1.

Magerus-Chatinet A, Stolzenberg MC, Loffredo M, Neven B, Schaffner C, Ducrot N, **Arkwright PD**, Bader-Meunier B, Barbot J, Blanche S, Casanova JL, Debré M, Ferster A, Fieschi C, Florkin B, Galumbrun C, Hermine O, Lambotte O, Solary E, Thomas C, Le Deist F, Picard C, Fischer A, Rieux-Laucat F. FAS-L, IL-10 and double-negative CD4-CD8-TCR alpha/beta+ T cells are reliable markers of ALPS associated with FAS loss of function. *Blood* 2009; 26:3027-30.

### 2008

Best J, **Hughes S**. Evidence behind the WHO Guidelines: hospital care for children--what are the useful clinical features of bacterial meningitis found in infants and children? *J Trop Pediatr*. 2008 54(2):83-6

Ryan KR, Hong M, **Arkwright PD**, Gennery AR, Costigan C, Domingues M, Denning D, McConnell V, **Cant AJ**, Abinun M, Spickett GP, Lilic D. Impaired Dendritic Cell maturation and Cytokine Production in Patients with Chronic Mucocutaneous Candidiasis with or without APECED. *Clin Exp Immunol* 2008;154:406-14

Meloni A, Furcas M, Cetani F, Marcocci C, Falomi A, Perniola R, Pura M, Boe Wolff A, Husebye E, Lilic D, Ryan KR, Gennery AR, **Cant AJ**, Abinun M, Spickett GP, **Arkwright PD**, Denning D, Costigan C, Dominguez M, McConnell V, Willcox N, Meager A. Autoantibodies against type I interferons: alternative diagnostic criteria for autoimmune polyendocrinopathy syndrome I (APS-I). *J Clin Endocrinol Metabol* 2008;93:4389-97..

Nedjai B, Hitman GA, Yousaf N, Chernajovsky Y, Stjernberg-Salmela S, Pettersson T, Ranki A, Hawkins PN, **Arkwright PD**, McDermott MF, Turner MD. Abnormal tumor necrosis factor receptor I cell surface expression and NF-kappaB activation in tumor necrosis factor receptor-associated periodic syndrome. *Arthritis Rheum* 2008;58:273-83.

## 2007

Vassallo G, Newton RW, Chieng SE, Haeney MR, **Arkwright PD**. Clinical variability and characteristic autoantibody profile in primary C1q complement deficiency. *Rheumatology* 2007;16:12-4.

**Arkwright PD**, Abinun M, **Cant AJ**. Mevalonic aciduria cured by bone marrow transplantation. *N Engl J Med* 2007;357:1350.

Balmer P, Borrow R, **Arkwright PD**. PPV-23 does not provide additional pneumococcal serotype antibody protection in children who have been given two doses of the PCV-7. *Vaccine* 2007;25:6321-5.

## 2006

West SL, Newton RW, Baildam EM, Turner AJ, **Arkwright PD**. Recurrent hemiplegia in an infant following third trimester maternal herpes zoster infection. *Dev Med Child Neurol* 2006;48:991-3.

Chapgier A, Boisson-Dupuis S, Jouanguy E, Vogt G, Feinberg J, Prochnicka-Chalufour A, Casrouge A, Yang K, Soudais C, Fieschi C, de Filipe Santos O, Bustamante J, Picardi C, de Beaucoudrey L, Emiles J-F, **Arkwright PD**, Schreiber RD, Rolinck-Werninghaus C, Rosen-Wolff A, Magdorfe K, Roesler J, Casanova J-L. Novel STAT1 alleles in otherwise healthy patients with mycobacterial disease. *PLOS genetics* 2006;2:e131.

Alachkar H, Taubenheim N, Haeney MR, Durandy A, **Arkwright PD**. Memory switched B-cell percentage and not serum immunoglobulin concentration is associated with clinical complications in children and adults with specific antibody deficiency and common variable immunodeficiency. *Clin Immunol* 2006;120:310-8.

Uddin S, Borrow R, Heaney MR, Moran A, Warrington R, Balmer P, **Arkwright PD**. Total and serotype-specific pneumococcal antibody titres in children and with normal immunity and immunodeficiency diseases. *Vaccine* 2006;24:5637-44.

Theilgaard-Mönch K, Jacobsen LC, Nielsen MJ, Rasmussen T, Udby L, Gharib M, **Arkwright PD**, Gombart AF, Calafat J, Moestrup SK, Porse BT, Borregaard N. Haptoglobin is synthesized during granulocytic differentiation, stored in specific granules, and released by neutrophils in response to activation. *Blood* 2006;108:353-61.

Chapgier A, **Wynn RF**, Jouanguy E, Filipe-Santos O, Zhang S, Feinberg J, Hawkins K, Casanova JL, **Arkwright PD**. Human complete Stat-1 deficiency is associated with defective type I and II interferon responses *in vitro* but immunity to some low virulence viruses *in vivo*. *J Immunol* 2006;176:5078-83.

**Wynn RF**, Sood M, Theilgaard-Mönch K, Jones C, Gombart AF, Gharib M, **Arkwright PD**. Severe and protracted diarrhoea of infancy caused by neutrophil specific granule deficiency and cured with stem cell transplantation. *GUT* 2006;55:292-6.

## 2005

Ryan KR, Lawson CA, Lorenzi AR, **Arkwright PD**, Isaacs JD, Lilic D. CD4+CD25+ T regulatory cells are decreased in patients with Autoimmune PolyEndocrinopathy Candidiasis Ectodermal Dystrophy. *J Allergy Clin Immunol* 2005;116:1158-9.

Theilgaard-Mönch K, Jacobsen LC, Rasmussen T, Niemann CU, Udby L, Borup R, Gharib M, **Arkwright PD**, Gombart AF, Calafat J, Porse BT, Borregaard N. Highly glycosylated alpha1-acid glycoprotein is synthesized in myelocytes, stored in secondary granules, and released by activated neutrophils. *J Leukocyte Biol.* 2005;78:462-70.

**Wynn RF**, **Arkwright PD**, Haque T, Gharib MI, Wilkie G, Morton-Jones M, Crawford DH. Allogeneic EBV-specific cytotoxic T lymphocyte infusions are effective in the treatment of EBV-driven primary CNS lymphoma. *Lancet Oncology* 2005;6:344-6.