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Immune abnormalities in Fontan protein-losing enteropathy and plastic bronchitis: a case control study

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Background

- Protein-losing enteropathy (PLE) and plastic bronchitis (PB) are complications of the Fontan palliation that carry significant morbidity and mortality
- Both diseases characterized by flares that are often associated with inflammatory processes including seasonal allergies and infections, suggesting that disease pathology may involve immune dysregulation
- Reports of Immune abnormalities in these diseases are described but limited by incomplete immune characterization and lack of a control group
- Study aims:
 - Fully characterize the quantitative immune status of Fontan patients with PLE or PB
 - Compare Fontans with PLE or PB to healthy control Fontans

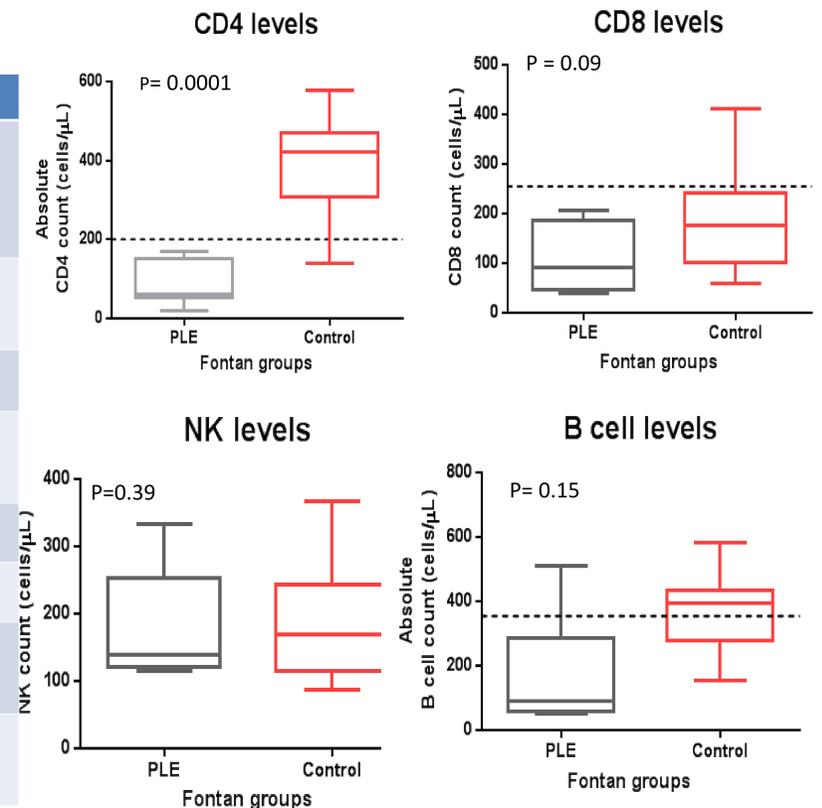
Methods

- Age-matched subjects with PLE or PB to "healthy" Fontan controls 1:1 or 1:2
- Prospectively obtained the following immunologic data:
 - Complete blood cell counts with differential, quantitation of lymphocyte subsets by flow cytometry, serum immunoglobulin and IgG subclass distribution, complement levels, total protein electrophoresis, and antibody titers to vaccines
- Two group comparisons utilized appropriate parametric and non-parametric tests. Continuous variables were categorized as low, normal, or high according to laboratory standards

Results

- A total of 20 patients were enrolled (6 PLE, 3 PB, 1 PLE and PB, 10 healthy control Fontans)
- Groups were similar in terms of underlying cardiac diagnosis, age at Fontan completion, length of stay after Fontan, incidence of thymectomy, vaccination history

	PLE	Control	P-value
Absolute Lymphocyte Count < 1.0	100%	29%	0.02
CD4 Count < 200 cells/ μ L	100%	0%	<0.001
Reversed CD4:CD8	71%		
Negative MMR response	71%	0%	0.01
Low IgG	100%	29%	0.01
Low IgA	57%	0%	0.01
Normal White Blood Cell Count	100%	100%	NS
Normal Complement	100%	100%	NS



- The PB group had only 3 patients and was likely underpowered, but was not significantly different from the control group

Conclusions

- Both healthy Fontans and patients with PLE or PB have quantitative immunological abnormalities
- In patients with PLE, more components of the immune system are affected with more severe derangements, including markedly depressed CD4 counts, and a high incidence of negative response to live vaccines
- Some of these abnormalities, in particular selectively decreased CD4 counts, cannot be explained simply by stool losses. To what extent these abnormalities cause, potentiate or support PLE, or lead to significant morbidities requires further study