CHD. 25 were simple or moderate complex non-cyanotic CHD. The estimated incidence was 1.828%. 17/47 (36.2%) were cyanotic. The incidence of simple CHD was 25/47(53.2%). Analysis of the specific anatomy is presented in table 1.

Abstract OC39 Table 1

1	BAov (functional/anatomic) no additional disease	3.035%
2	VSD (isolated/all types)	14.9%
3	ASD (isolated/all types)	8.5%
4	PDA (isolated)	6.384%
5	c-AVSD	4.258%
6	A-P window	2.128%
7	Coronary artery abnormalities	4.256%
8	PA valve stenosis	4.256%
9	Ao valve stenosis	4.256%
10	Mitral valve disease	2.128%
11	CoA (isolated/all types)	4.256%
12	IAA (isolated/all types)	2.128%
13	ToF (all types)	6.384%
14	d-TGA (all types)	4.256%
15	PAv atresia with IVS	4.256%
16	DORV (all types)	4.256%
17	TAPVD (all types)	4.256%
18	Tricuspid valve atresia	2.128%
19	HLHS	2.128%
20	c-c-TGA	2.128%
21	Truncus Arteriosus	2.128%
22	Ebstein Anomaly	2.128%
23	Univentricular Anatomy	2.128%
24	Heterotaxia Syndromes	2.128%
25	Shone's Complex	2.128%
26	Core triatriatum	2.128%

Conclusions This first-ever prospective epidemiology study in Azerbaijan involving a cohort equal to 1.65% of the annual living births of the country, estimated a high incidence. This is among the highest reported globally. The amount of critical & cyanotic CHD was 46.8% and 36.2% respectable. These high numbers are possibly related to an isolated population and conjugated marriage customs of the country. This represents a public population health burden.

OC40

IS DOWN SYNDROME RELATED ARTHRITIS (DA) A DISTINCT DISEASE FROM JUVENILE IDIOPATHIC ARTHRITIS (JIA)?

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DA is frequently polyarticular-RF-negative with predominance in the small joints of the hands and wrists. It has an erosive phenotype, and clinically appears to be a more aggressive disease than JIA. Arthritis occurs more frequently in children with Down syndrome (DS), with prevalence 20 times greater than JIA. To date little is known about the underlying mechanisms that drive DA disease pathogenesis, however these features suggest that DA may be distinct from JIA. We wanted to explore this hypothesis by examining B-cell subsets and T-

cell cytokine profiles; and characterising and comparing the synovial membrane immunohistochemistry in DA versus JIA.

Methods Multicolour-flow cytometry was used to analyse B and T cell phenotypes in PBMCs from 40 children (n=10/group - Healthy Control (HC), JIA, DS, DA), and assessed by Flowjo software analysis. Cells were stained as follows; B-cell panel (CD38, CD24, CD20, CD80, CD27, IgM, CD138, CD45, CD19, MHC class II, BCMA, CD40, CD86, IgD); T-cell panel - cytokines analysed after 5-hours PMA/Ionomycin stimulation (CD3, CD8, CD161, IFN- γ , TNF- α , IL-17a, GM-CSF).

Synovial tissue was obtained through US-guided biopsy and analysed by immunohistochemistry for CD3, CD20, CD68, FVIII (DA n=3; JIA n=4). Levels of vascularity and lining layer hyperplasia were also scored. Analysis was performed using a semi-quantification scoring method.

Results Flow cytometry analysis revealed that children with DA had a significantly lower number of circulating CD19 +CD20+ B cells when compared to children with JIA ($p<0.05$) and HC ($p<0.001$). However, they had a greater proportion of memory B cells (CD27+) when compared to children with DS ($p<0.05$).

IFN- γ and TNF- α production by CD8+/CD8- T cells was greater in DA compared to both JIA (CD8+IFN γ + $p<0.001$; CD8+TNF α + $p<0.01$; CD8-IFN γ + $p<0.05$; CD8-TNF α $p<0.05$) and HC (CD8+IFN γ + $p<0.05$; CD8+TNF α + $p<0.05$; CD8-IFN γ + $p<0.05$; CD8-TNF α $p<0.01$).

Examination of synovial tissue demonstrated marked increase in synovial lining layer hyperplasia in DA (median 6 (3-9)) versus JIA (median 3(2-4)). Higher levels of CD3+ cells ($p<0.05$), Macrophages ($p<0.05$), CD20+ cells and FVIII in DA were also observed.

Conclusion Significant differences were observed in the immune and histological profiles of DA and JIA. These differences may begin to explain the erosive phenotype observed in DA. This study is important as it is the first of its kind to describe the immuno-histology of DA. Understanding the pathogenesis of DA will allow for more targeted, personalised treatment approaches, in turn improving outcomes and therefore quality of life for children with Down syndrome and arthritis.

OC42

PHARMACOLOGICAL INDUCTION OF A20: ANTI-INFLAMMATORY EFFECTS IN PAEDIATRIC ASTHMA

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Background Chronic exaggerated inflammation in asthma is mediated by the activation of pro-inflammatory transcription factor NF- κ B(p65) and an intrinsic lack of its down-regulator A20. A20 has strong anti-inflammatory effects when induced, and as such could be an attractive anti-inflammatory pharmacological target. Bioinformatic approaches using statistically significant connection Map (sscMap)) predict that prednisone and amitriptyline would induce A20 in asthmatic airway epithelial cells. We performed a proof of concept study to determine whether amitriptyline applied to nasal airway epithelial cells (AEC) from children could be beneficial in asthma.