Pediatric Antiphospholipid Syndrome

Antiphospholipid syndrome (APS) is an uncommon entity in childhood. The entity was described for the first time in early 1980s and the antibodies implicated were anti-cardiolipin antibodies. About half of children with APS have primary APS. In children, APS is more commonly associated with SLE than in adults. Association of various manifestations of systemic lupus erythematous (SLE) were found to be associated with the presence of anti-phospholipid antibodies. The common ones being portal and pulmonary hypertension (1), cerebral infarction (2), thrombocytopenia (3), pulmonary hypertension (4) and chorea (5). Diagnosis of APS including paediatric APS is based upon clinical features of either vascular thrombosis or pregnancy morbidity in the presence of antiphospholipid antibodies. It is obvious that pregnancy morbidity is not of relevance in paediatric age group and there are other clinical features like lived reticularis, chorea that suggest the presence of APS in children. These cutaneous, neurological and haematological manifestations are not taken into account while defining APS in children. Evidence- based recommendations for diagnosis and treatment of paediatric APS have been recently published by The Single Hub and Access point for paediatric Rheumatology in Europe (SHARE) (6).

73 children with SLE were tested for anti-phospholipid antibodies. 56 (76.7%) were girls. 7 had LAC (DRVV) positivity, only one had LAC (SCT) positivity. Only 2 had LAC (DRVV) positivity on repeat testing. The one who had LAC (SCT) positivity turned negative on repeat testing, but a new patient tested positive at repeat testing for LAC (SCT). 20 had ACA IgM positivity, 3 had ACA IgG positivity at the time of initial testing. ACA IgG was not detected in any of the patients while ACA IgM was detected in 7 on repeat testing. Anti-β2GP1 was not detected in any of the patients on initial testing but Anti-β2GP1 IgG was found in one on repeat testing. One patient had triple positivity (on repeat testing). Six patients had thrombotic complications with SLE. Two had pulmonary thromboembolism (PTE), 1 had deep vein thrombosis (DVT) of lower limbs, 2 patients had old CNS infarcts and 1 had acute infarct in CNS. One patient with LA hypoprothrombinemia syndrome presented with refractory coagulopathy and bleeding.

	LAC DRVV	LAC SCT	ACA IgG	ACA IgM	Aβ2GP1 IgG	Aβ2GP1 IgM
Initial	7	1	3	20	0	0
Repeat	2	**1**	0	7	1	0

Clinical manifestation	**APLs**	**Repeat APLs**
PTE	ACA IgM	Negative
PTE	Negative	Negative
DVT	ACA IgM	ACA IgM
Parietal lobe infarct	LAC, ACA IgM	Negative
Old MCA infarct	ACA IgG	Negative
Acute CNS infarct	ACA IgM	Negative
Coagulopathy	LAC	Pending

1. Mackworth-Young CG, Gharavi AE, Boey ML, Hughes GRV. **Portal and pulmonary hypertension** in a case of SLE: possible relationships with clotting abnormality. *Eur J Rheumtol Inflamm* 1984;7:72-74.

2. Harris EN, Gharavi AE, Asherson RA, Boey ML, Hughes GR. **Cerebral infarction** in SLE. Association with anticardiolipin antibodies. *Clin Exp Rheumatol* 1984;2:47-51.

3. Harris EN, Gharavi AE, Hegde U, Hughes GRV. Anticardiolipin antibodies in **autoimmune thrombocytopenic purpura**. *Br J Haematol* 1985;159:231-234.

4. Asherson RA, Hackett D, Gharavi AE, Harris EN, Kennedy HG, Hughes GR. **Pulmonary hypertension** in SLE: a report of 3 cases. *J Rheumatol* 12:416-418

5. Asherson RA, Hughes GRV. Antiphospholipid antibodies **and chorea**. *J Rheumatol* 1987;15:377-379.

6. Groot N, de Graeff N, Avcin T, Bader-Meunier B, Dolezalova P, Feldman B, Kenet G, Koné-Paut I, Lahdenne P, Marks SD, McCann L, Pilkington CA, Ravelli A, van Royen-Kerkhof A, Uziel Y, Vastert SJ, Wulffraat NM, Ozen S, Brogan P, Kamphuis S, Beresford MW. European evidence-based recommendations for diagnosis and treatment of paediatric antiphospholipid syndrome: the SHARE initiative. Ann Rheum Dis. 2017 May 4