Results: As a result of genetic research performed due to recurrent infection and low immunoglobulin, he was diagnosed with immunodeficiency 36 with lymphoproliferation (MIM: 616005). The patient is still receiving anticoagulation with warfarin and Ig replacement therapy every 3 weeks.
Conclusion(s): The development of immune deficiency and macrophage activation syndrome as the final diagnosis during follow-up in a patient presenting with multiple deep venous thrombosis is a rare clinical presentation. We believe that it is important to investigate other underlying diseases in multiple thrombosis.

PB0592

Antiphospholipid Syndrome in Children

C. Albayrak
Manager of hemophilia center, Samsun, Samsun, Turkey

Background: Antiphospholipid syndrome(APS) is a rare autoimmune disease of unknown etiology that represents the leading cause of acquired thromboembolism and recurrent miscarriage. It is characterized by the persistently elevated presence of pathogenic antiphospholipid autoantibodies against cardiolipin, ß2-glycoprotein-I and/or a positive lupus anticoagulant test. Although it is more common in adults, APS can sometimes occur in the neonatal period and childhood. Adult-onset classification criteria have poor validity for the pediatric population (where pregnancy-related complications are rare) and, as a result, the prevalence of the disease in childhood is difficult to assess. Thromboembolic events in children include stroke and pulmonary embolism, as well as deep vein thrombosis, which can cause serious long-term disability.
Aims: Our aim in this study is to retrospectively evaluate pediatric APS cases diagnosed in our clinic.
Methods: Eight pediatric patients were diagnosed with APS in our clinic. Six of the patients were girls and two were boys. Ages at diagnosis were between 10 and 16.
Results: All patients were diagnosed with APS secondary to SLE. Presentation findings were skin rash, thrombocytopenia, involuntary movements(chorea) or thromboembolic events. All patients were diagnosed in the pediatric hematology department and were followed up in the rheumatology department due to the underlying diagnosis of SLE. Follow-up periods vary between 1-20 years. All patients are receiving anticoagulant treatment. Thromboembolic events have been observed when treatment compliance is disrupted.
Conclusion(s): Pediatric APS cases present with hematological findings and are diagnosed by pediatric hematologists. Diagnosis of pediatric APS is more difficult because diagnostic criteria were developed for adults. Since pregnancy morbidity does not usually occur in children, it is difficult to complete the diagnostic criteria. Diagnostic criteria for pediatric APS cases should be redefined. Therefore, we believe that it would be beneficial to collect and evaluate cases retrospectively and prospectively in a multicenter manner for this rare disease.

PB0591

Differences in Types of Thromboses and Risk Factors in Males versus Females in Adolescent Thromboembolism in the Throm-PED Registry

L. Srivaths[1], N. Montanez[2] and H. Van Ommen[3]
[1]*University of Texas Health Science Center of Houston, McGovern Medical School, Gulf States Hemophilia and Thrombophilia Center, Houston, Texas, United States,* [2]*University of Texas Health Science Center of Houston, McGovern Medical School, Department of Pediatrics, Division of Hematology, Gulf States Hemophilia and Thrombophilia Center, Houston, Texas, United States,* [3]*Erasmus University Medical Center, Sophia Children's Hospital Erasmus MC, Rotterdam, The Netherlands, Rotterdam, Zuid-Holland, Netherlands*

Background: Adolescents are at heightened risk for thromboembolism (TE), likely due to diagnostic/therapeutic advances, and unique health challenges. Varying risk factors in females compared to males (e.g., estrogen, autoimmune disorders/antiphospholipid syndrome) may result in differences in prevalence, and TE types, which in turn can lead to differences in treatment choices, outcome and complications. Few studies have evaluated sex-based differences in TE and no study has addressed the impact of sex in adolescent TE.
Aims: Evaluation of sex-based differences in adolescent TE characteristics and management.
Methods: The Throm-PED registry is an international, multicenter, prospective registry of the International Pediatric Thrombosis Network (IPTN). Following data prospectively collected in Throm-PED registry (overall-since 2019; specific adolescent TE protocol-since 2021) were reviewed: age at diagnosis, sex, TE type, location, risk factors, treatment.
Results: Data were analyzed for 776 adolescents with TE, M-360 (46%), F-416 (54%), median age 15 years (range-10-21 years) in both sexes (Table 1,2). There was statistically significant greater prevalence of arterial thromboses overall, and intracranial venous, intracardiac, and aortic thromboses in males, and venous TE overall, and pulmonary embolism in females. For risk factors, congenital heart disease, surgery, and previous thrombotic event in males and oral contraceptives, obesity, antiphospholipid syndrome, and autoimmune disorders in females, were more prevalent which were statistically significant. Therapeutic modalities were similar in both.
Conclusion(s): Sex-based differences appear to impact adolescent TE characteristics with higher female prevalence as noted in our study. Increased prevalence of congenital heart disease may explain the increased prevalence of arterial/intracardiac/aortic thromboses in males. Risk factors with higher prevalence in females including estrogen therapy, autoimmune disorders, antiphospholipid syndrome and obesity might have contributed to increased prevalence of venous TE/pulmonary embolism when compared to males. Future analysis with continued follow-up we hope will shed light on the sex-based differences in TE outcome and complications.

Table 1. Adolescent Thromboembolism Baseline Patient Characteristics in Males versus Females

Patient Characteristics	Gender		
	Male N (%)	Female N (%)	p value (Chi-square)
Total N=776 (100%)	360 (46%)	416 (54%)	
Age (years)			
Median	15	15	
Range	10-21	10-21	
Type of Thrombosis			
Venous	327 (90%)	405 (97%)	p=0.0001
Arterial	40 (11%)	13 (3%)	p=0.0001
Location: Venous Thromboembolism			
Upper Extremity	68 (19%)	97 (23%)	P=0.13
Subclavian Vein	15 (4%)	20 (5%)	P=0.60
Jugular Vein	25 (7%)	40 (10%)	P=0.18
Lower Extremity	106 (29%)	121 (29%)	P=0.9
Inferior Caval Vein	9 (3%)	14 (3%)	P=0.47
Superior Caval Vein	5 (1%)	9 (2%)	P=0.41
Abdominal Vein	32 (8%)	29 (7%)	P=0.32
Intracranial	58 (16%)	37 (9%)	P=0.003
Pulmonary Embolism	63 (18%)	102 (25%)	P=0.018
Other	12 (3%)	23 (6%)	P=0.14
Location: Arterial Thrombosis			
Upper Extremity	1 (0.02%)	1 (0.002%)	P=0.9
Lower Extremity	6 (2%)	2 (0.7%)	P=0.15
Intracardiac	23 (6%)	6 (1%)	P=0.001
Aorta	6 (2%)	0	P=0.01
Other	5 (1%)	5 (1%)	P=0.8

Table 2. Adolescent Thromboembolism Risk Factors and Treatment in Males versus Females

Patient Characteristics	Gender		p value
	Male N (%)	Female N (%)	(Chi-square)
Risk Factors			
None	28 (8%)	35 (8%)	P=0.7
Central Venous Catheter	93 (26%)	99 (24%)	P=0.5
Congenital Heart Disease	23 (6%)	10 (2%)	**P=0.006**
Oral Contraceptives	0 (0%)	94 (23%)	**P=0.0001**
Malignancy	55 (15%)	64 (15%)	P=0.9
Infection	78 (22%)	82 (20%)	P=0.5
COVID-19 infection	15 (4%)	21 (5%)	P=0.5
Surgery	55 (15%)	42 (10%)	**P=0.03**
Immobility	70 (19%)	67 (16%)	P=0.5
Renal Disease	12 (3%)	20 (5%)	P=0.3
Sickle Cell Disease	4 (1%)	3 (0.7%)	P=0.5
Obesity	31 (9%)	58 (14%)	**P=0.024**
Arterial Catheter	1 (0.02%)	5 (1%)	P=0.2
Previous Thrombotic Event	37 (10%)	25 (6%)	**P=0.03**
Congenital Thrombophilia	28 (8%)	30 (7%)	P=0.7
Acquired Thrombophilia	4 (1%)	5 (1%)	P=0.8
Antiphospholipid Syndrome	4 (1%)	15 (4%)	**P=0.03**
Autoimmune Disorder	3 (0.8%)	20 (5%)	**P=0.001**
Positive Family Hx	21 (6%)	23 (6%)	P=0.8
Anatomic Abnormalities	15 (4%)	21 (5%)	P=0.6
Smoking	2 (0.5%)	1 (0.2%)	P=0.5
Trauma	18 (5%)	14 (3%)	P=0.3
Other	89 (25%)	88 (21%)	P=0.26
Treatment: Medical			
None	8 (2%)	12 (3%)	P=0.5
Antiplatelet Drugs	11 (3%)	8 (2%)	P=0.35
Unfractionated Heparin	93 (26%)	119 (29%)	P=0.4
LMWH	276 (77%)	300 (72%)	P=0.16
Enoxaparin	243 (68%)	259 (62%)	P=0.13
Other (Dalteparin, Nadroparin, Tinzaparin)	32 (9%)	43 (10%)	P=0.5
Vitamin K Antagonist	24 (7%)	42 (10%)	P=0.09
Warfarin	20 (6%)	31 (7%)	P=0.3
Acenocoumarol/Phenprocoumon	6 (2%)	15 (4%)	P=0.12
Fondaparinux	4 (1%)	7 (2%)	P=0.5
Bivalirudin	6 (2%)	2 (0.5%)	P=0.15
DOAC	113 (31%)	150 (36%)	P=0.17
Rivaroxaban	103 (29%)	124 (30%)	P=0.7
Dabigatran	5 (1%)	8 (2%)	P=0.7
Apixaban	19 (5%)	31 (7%)	P=0.24
Edoxaban	1 (0.02%)	1 (0.2%)	P=0.9
Other	7 (2%)	6 (1%)	P=0.59
Treatment: Interventional			
Thrombolysis	18 (5%)	27 (6%)	P=0.4
Mechanical Intervention	6 (2%)	5 (1%)	P=0.7
IVC Filter	0	1 (0.2%)	P=1.0
Treatment: Surgical	4 (1%)	4 (1%)	P=1.0

PB0590

Venous Thrombo-embolism in Transgender and Gender Non-Binary Adolescents: An Administrative Database Study

R. Kumar[1], C. Baskaran[2], S. Roberts[2], N. Chen[3] and W. London[1]

[1]*Dana Farber/Boston Children's Cancer and Blood Disorders Center; Harvard Medical School, Boston, Massachusetts, United States,* [2]*Boston Children's Hospital; Harvard Medical School, Boston, Massachusetts, United States,* [3]*Dana Farber/Boston Children's Cancer and Blood Disorders Center, Boston, Massachusetts, United States*

Background: A recent study conducted at Boston Children's Hospital (BCH) showed that venous thromboembolism (VTE) is rare in transgender and gender non-binary (TNB) adolescents. Additionally, gender-affirming hormone-treatment (GAHT) was not found to be a significant risk factor for thrombosis in this cohort.

Aims: The principal objective of this investigation was to confirm our preliminary findings across 49 tertiary-care children's hospitals in the United States (US).

Methods: Data were obtained from Pediatric Health Information System (PHIS), an administrative database of clinical/resource-utilization data from 49 tertiary-care children's hospitals in the US. ICD9/ICD10 codes were used to identify eligible subjects, defined as patients (birth-21 years) hospitalized with VTE between 1/1/2012-12/31/2021 at one of the 49-participating centers. ICD9/ICD10 codes for gender dysphoria were used to identify TNB patients. Mean (SD) or median (range) were calculated for continuous data, and frequency with 95% confidence interval for categorical data.

Results: During the study period, 39,503 unique patients were diagnosed with VTE. Mean age (SD) at VTE diagnosis was 7.7 (7.3) years. Only 23 (0.06%) patients had a diagnosis code for gender dysphoria. Among these 23 patients, the median (range) age at VTE diagnosis was 16.2 (11.6-20.5) years. Compared to the overall cohort, TNB patients with VTE were more likely to be female, white race, and/or obese (Table 1). Only 2/23 (8.7%) TNB patients with VTE had an ICD9/ICD10 code for GAHT. We have sought approval from the Institutional Review Board to de-identify patients from BCH enrolled in PHIS for internal-validation of these data.

Conclusion(s): These findings support our initial observation that VTE is rare in TNB adolescents, and typically occurs in the setting of additional risk-factors. Only 2/23 TNB patients had an ICD9/ICD10 code for GAHT, suggesting that it is unlikely that GAHT is a significant risk factor for VTE in this cohort. Internal-validation of these data is currently underway.

	All VTE patients (n=39,503)		Subset of VTE patients were TNB (n=23)	
Characteristic	**n**	**mean (SD)**	**n**	**median (range)**
Age at VTE diagnosis (years)	39,503	7.7 (7.3)	23	16.2 (11.6, 20.5)
	n	% (95% CI)	n	% (95% CI)
Birth gender				
Female	18,395	47% (46%, 47%)	17	74% (52%, 90%)
Male	21,092	53% (53%, 54%)	6	26% (10%, 48%)
Race				
White	23,546	63% (62%, 63%)	18	82% (60%, 95%)
Black	7,362	20% (19%, 20%)	1	5% (0.12%, 23%)
Asian	1,173	3% (3%, 3.3%)	0	0% (0%, 15%)
Other	5,413	14% (14%, 15%)	3	14% (2.9%, 35%)
Unknown	2,009		1	
Ethnicity				
Hispanic	6,951	19% (19%, 20%)	2	10% (1.2%, 32%)
Non-Hispanic	29,332	81% (80%, 81%)	18	90% (68%, 99%)
Unknown	3,220		3	
Venous thrombosis diagnosis				
Lower extremities	10,754	27% (27%,28%)	8	35% (16%, 57%)
Upper extremities	11,894	30% (30%,31%)	5	22% (7.5%, 44%)
Pulmonary embolism	3,894	9.7% (9.5,10%)	5	22% (7.5%, 44%)
CSVT	980	2.5% (2.3,2.6%)	1	4% (0.11%, 22%)
Other	12.026	30% (30%,31%)	4	17% (5.0%, 39%)
Mortality				
Death during hospitalization	2,790	7% (6.8%, 7.3%)	0	0% (0%, 15%)
Comorbid conditions associated with VTE				
Congenital Heart Disease				
Yes	11,773	30% (29%, 30%)	0	0% (0%, 15%)
Inflammatory bowel disease				
Yes	460	1% (1.1%, 1.3%)	1	4% (0.11%, 22%)
Chronic renal disease				
Yes	1,983	5% (4.8%, 5.2%)	3	13% (2.8%, 34%)
Cancer				
Yes	4,188	11% (10%, 11%)	2	9% (1.1%, 28%)
Obesity				
Yes	2,871	7% (7%, 7.5%)	10	43% (23%, 66%)

Table 1: Characteristics of patients diagnosed with VTE between 2012–2021 (n=39,503), compared to a subset who were identified as TNB

Neonatal Coagulation Disorders

PB0602

Compound Heterozygous congenital Protein C deficiency: Report of a challenging case with recurrent purpura fulminans treated with Protein C concentrations: Case Report

C. Srichumpuang[1] and D. Sosothikul[2]

[1]*King Chulalongkorn Memorial Hospital, Bangkok, Krung Thep, Thailand,* [2]*Chulalongkorn University, Pathumwan, Krung Thep, Thailand*

Background: Congenital protein C deficiency is a rare inherited disorder caused by alterations in the protein C gene (PROC) associated with an increased risk of developing thrombosis. Patients with biallelic PROC variants have more severe symptoms, typically causing purpura fulminans with severe venous thromboembolism in the neonatal period.