

**Supplementary table 1:** Special circumstances in specific paediatric liver diseases with distinct impact on haemostasis; a summary of data extracted from mainly small paediatric studies and case reports.

Diagnosis	Specific mechanism with distinct impact on haemostasis	Haemostatic laboratory assays	Reported bleeding complications	Reported thrombotic events/prothrombotic conditions	Reference
<b><i>Abnormal development of the hepatobiliary system</i></b>					
Alagille syndrome	Vascular anomalies		Yes. Intracranial bleeding.	Yes. Ischemic stroke	1 2
Biliary Atresia	Vitamin K malabsorption	INR↑, Factor II (FII), FVII, FIX, FX, Protein S and Protein C↓	Yes. Intracranial bleeding. Variceal bleeding.	Yes. Portal vein thrombosis	3-5
Elevated bile acids		Coagulation factors↑			6
<b><i>Inborn errors of metabolism</i></b>					
Artrogryposis-renal-cholestasis (ARC)	Absent or defect platelet granulae		Yes. Related to liver biopsy.		7-9
Carbohydrate deficient glycoprotein syndrome (CDG) type 1-2	Glycosylation defects	Coagulopathy, both coagulation factors and inhibitors ↓	Yes	Yes. Stroke-like episodes. Deep vein thrombosis. Disseminated intravascular coagulation (DIC).	10 11
Cystic fibrosis	Inflammation/infection. Antiphospholipid antibodies. Platelet hyperreactivity.			Yes. Especially related to central venous catheters (CVC).	12
Galactosemia	Glycosylation defects	Coagulopathy, both coagulation factors and inhibitors ↓	Yes. Vitreous haemorrhage.	Yes. Purpura Fulminans	13-15
Gauchers disease, type 3	Acquired vonWillebrand's disease. Platelet dysfunction	Coagulation factors↓ Platelets ↓ Fibrinolytic markers↑	Yes, Mucocutaneous bleeding. Muscular bleeding. Post-operative bleeding.		16 17
Glycogen storage disease type 1 ( von Gierkes disease)	Acquired vonWillebrand's disease. Platelet dysfunction	Coagulation factors↑ Inhibitors↑	Yes. Mucocutaneous bleeding. Peri-operative bleeding.		18-20
Tyrosinemia		Severe coagulopathy (in combination with α-fetoprotein ↑↑).	Yes, Cutaneous bleeding. Lung haemorrhage. Post-operative bleeding.		21 22

<b><i>Immune mediated disorders</i></b>					
Autoimmune hepatitis	Antiphospholipid antibodies				23
Gestational alloimmune liver disease (GALD) - Neonatal Haemochromatosis		Coagulopathy presenting sign, (in combination with glucose↓). Severe thrombocytopenia.	Yes. Pulmonary haemorrhage. Intracranial haemorrhage	Yes. Multi organ failure	24 25
Haemophagocytic lymphohistiocytosis	Hyperfibrinolysis. Platelet defects.	Low fibrinogen Thrombocytopenia	Yes, i.e. petechiae. mucocutaneous bleeding, bleeding after invasive procedures, pulmonary haemorrhage.		26-28
<b><i>Infections</i></b>					
Hepatitis A	Antiphospholipid antibodies			Yes. Budd-Chiari syndrome	29
Liver abscess	Endothelitis, inflammation, infection.			Yes. Portal vein thrombosis. Inferior vena cava thrombosis.	30 31
<b><i>Xenobiotic-induced liver injury</i></b>					
Venoocclusive disease	Disruption of sinusoidal endothelial lining and fibrin disposition in sinusoids and veins.		Yes. Liver-biopsy related	Yes. Microthrombosis and multi-organ failure	32
<b><i>Vascular disorders</i></b>					
Portosystemic shunt		Coagulopathy, both coagulation factors and inhibitors ↓			33
<b><i>Malignant disease</i></b>					
Hepatoblastoma			Yes. Post-operative bleeding	Yes. Post-operative thrombosis	34

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