Case Study: Unusual Course of the Hemolytic Disease of Fetus and Newborn (HDFN)

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Outline of the presentation

- Introduction of the case
- HDFN: etiology, pathophysiology, diagnosis and treatment
- Clinical course of the case
- Conclusion

Initial presentation

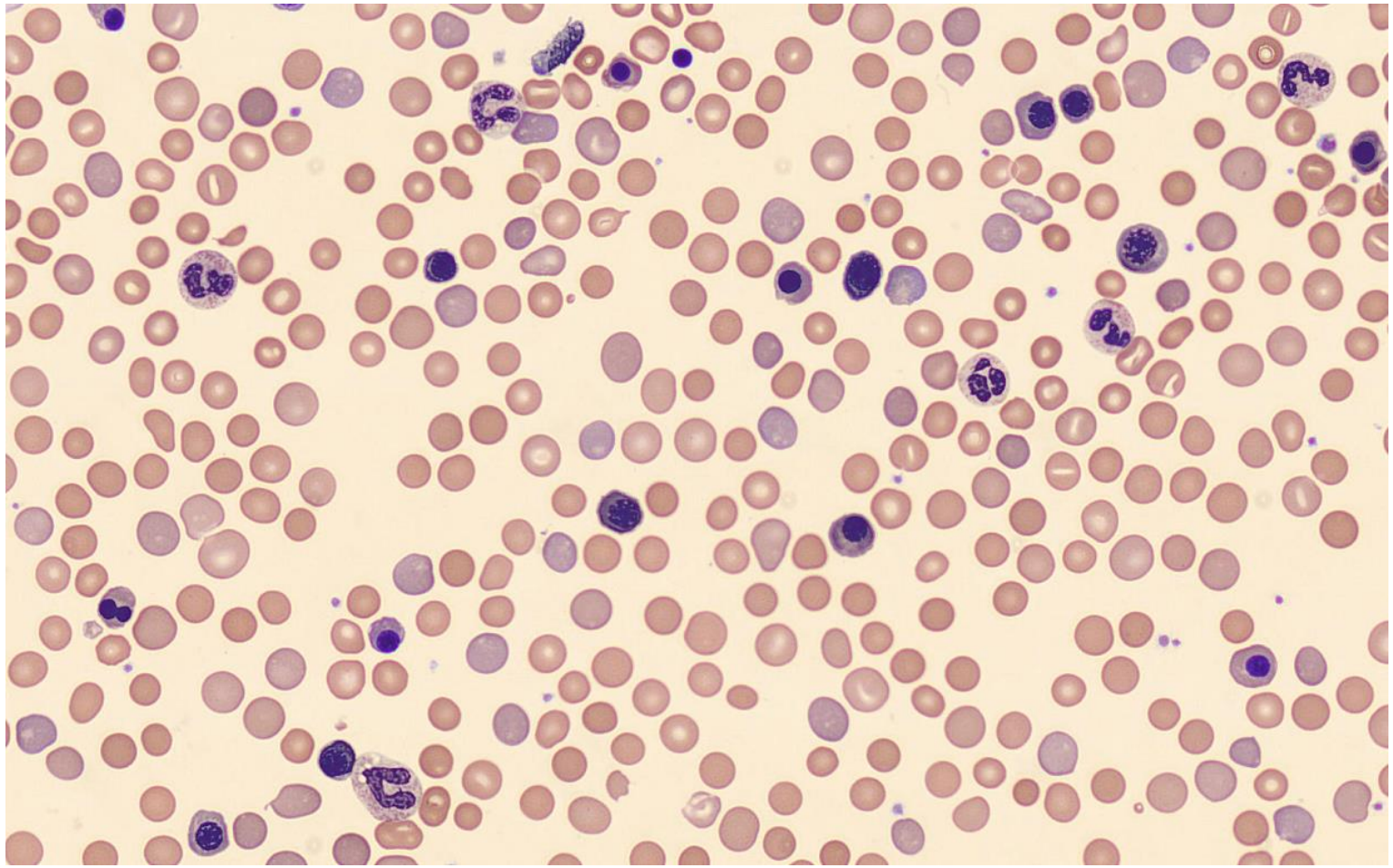
- Patient: 1-day old/37 weeks of gestation/3.3 kg infant
- Mother: G4P3; no prenatal care, no past medical history of HDFN
- Labor History: C-section
  - Apgar score: 3-3-6
  - Respiratory failure, C-pap

Initial Laboratory Markers

- Blood gases: pH 7.18; pCO2 66.9
- CBC:
  - 24/6.8/21%/120K
  - MCV> 130; MCH 54 and MCHC 31
  - ARC: 0.22 (16:00), next day 0.635
  - Abs. nucleated RBCs> 20
  - Giant platelets present
- Blood Bank testing:
  - O positive
  - Antibody screen: 0/3+/2+
  - DAT (IgG): 4+
  - Antibody identification: Anti E and anti c

- Chemistry:
  - BUN/Cr: 4/0.85
  - TB: 12.8, mainly indirect
  - AST/ALT: 225/39
  - Lactic Acid: 2.3
  - Na: 131, Calcium 7.9
- Coagulation:
  - PT/INR/aPTT: 18/1.4/35
  - Fibrinogen 156
  - DD: 8.2

Peripheral Smear



Diagnosis

- Severe hemolytic disease of fetus/newborn (HDFN)

What is HDFN?

- HDFN occurs when the blood types of a mother and baby are incompatible.
- 4000 cases/year
- Maternal alloantibodies are due to previous pregnancy, transfusion, HPSC transplantation.
- It is more likely to happen during second or subsequent pregnancy.

HDN Classification

ABO Incompatibility

- Group A infants/group O mothers of Eu or Asian ancestry
- Group B infants/group O mothers of African ancestry
- Rarely, severe HDFN

Rh Incompatibility

- 3x more common in Caucasian vs AA infants

Non-Rh Incompatibility

- Kell alloantibodies
- Fya alloantibodies
- Other alloantibodies: Fyb, Jka, Jkb, S, s

Pathophysiology of the HDFN

Mother

- Red cell antibodies
- IgM and IgG

Fetus/Newborn

- Antigen + RBCs
- Hemolysis
- Anemia with erythroblastosis
- Elevated bilirubin (total and indirect)
- Hydrops fetalis/Heart failure
- Extramedullary erythropoiesis
  - Hepatomegaly
  - Splenomegaly

Diagnosis & Treatment

- Pregnancy
- Type and screen
- Preventable disease: Rhogam
- Ultrasound
- Amniocentesis

- After birth diagnosis
- Complete blood count
- Type and screen

- Treatment during pregnancy
- Intrauterine transfusion
- Early delivery of the baby

- After birth treatment
- Phototherapy
- IV fluids
- RBC transfusion
- Intravenous immunoglobulin (IVIG)
- Blood exchange

Prenatal Care

Maternal antibody titers above critical value or previously affected pregnancy

Obtain paternal zygosity and antigen phenotype

Father homozygous-Use MCA Doppler US-Fetal blood sampling

Father heterozygous or not available-Cell free fetal DNA testing or amniocentesis : Fetus Ag+-MCA Doppler US or Fetus Ag—Routine prenatal care

Father negative for antigen-Routine prenatal care

Possible complications, lifelong considerations, future pregnancies

- Hyperbilirubinemia
- Brain damage
- Hearing damage
- Heart failure
- HDN is a short-term condition.
- The antibodies are usually gone by 8-12 weeks of life.
- Future pregnancies might be affected.

Going back to our case……

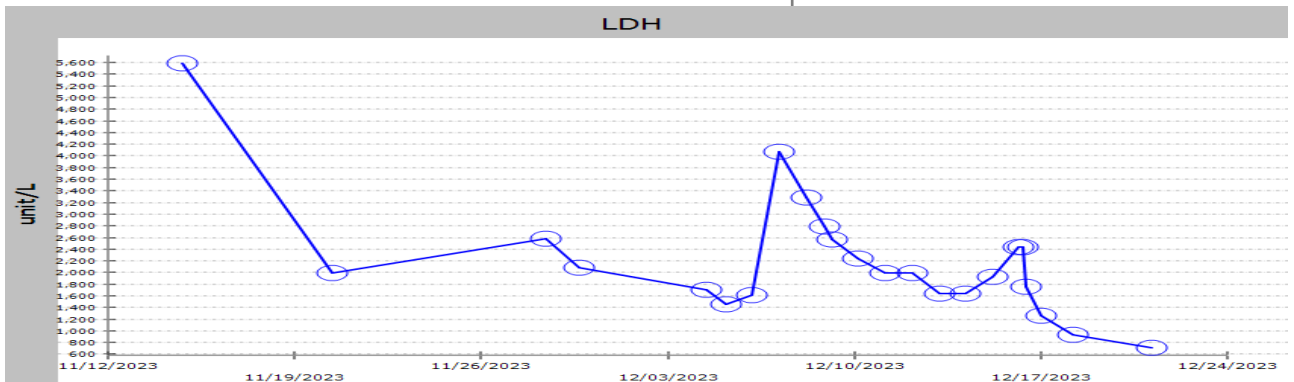
Diagnosis and Treatment

Day 1-2

- Severe Hemolytic Disease of fetus/newborn (HDFN)
- Heart failure
- Respiratory distress

- LDH> 5000
- Hemodynamically unstable

- Photo therapy
- RBC transfusions
- IVIG
- Orders for exchange
- Extracorporeal membranous oxygenation (ECMO) cannulation
- Therapeutic plasma exchange (TPE)



Day 4-7

- Generalized edema
- Abdominal distension
  - Hepatosplenomegaly
  - Hemorrhagic ascites

- ECMO
- Photo therapy
- Continuous renal replacement therapy (CRRT) initiated
- Paracentesis (abdominal tap)

Days 8-12

- Photo therapy associated rash
- Testing for porphyria triggered

- Phototherapy discontinued
- TPE initiated due to elevated plasma free hemoglobin (PFH) levels
- Seven daily TPE procedures performed in tandem with ECMO and CRRT

Day 19-22

TPE procedures completed

ECMO- patient was decannulated

CRRT support-continued

Eculizumab

Day 24

Total bilirubin level: 21

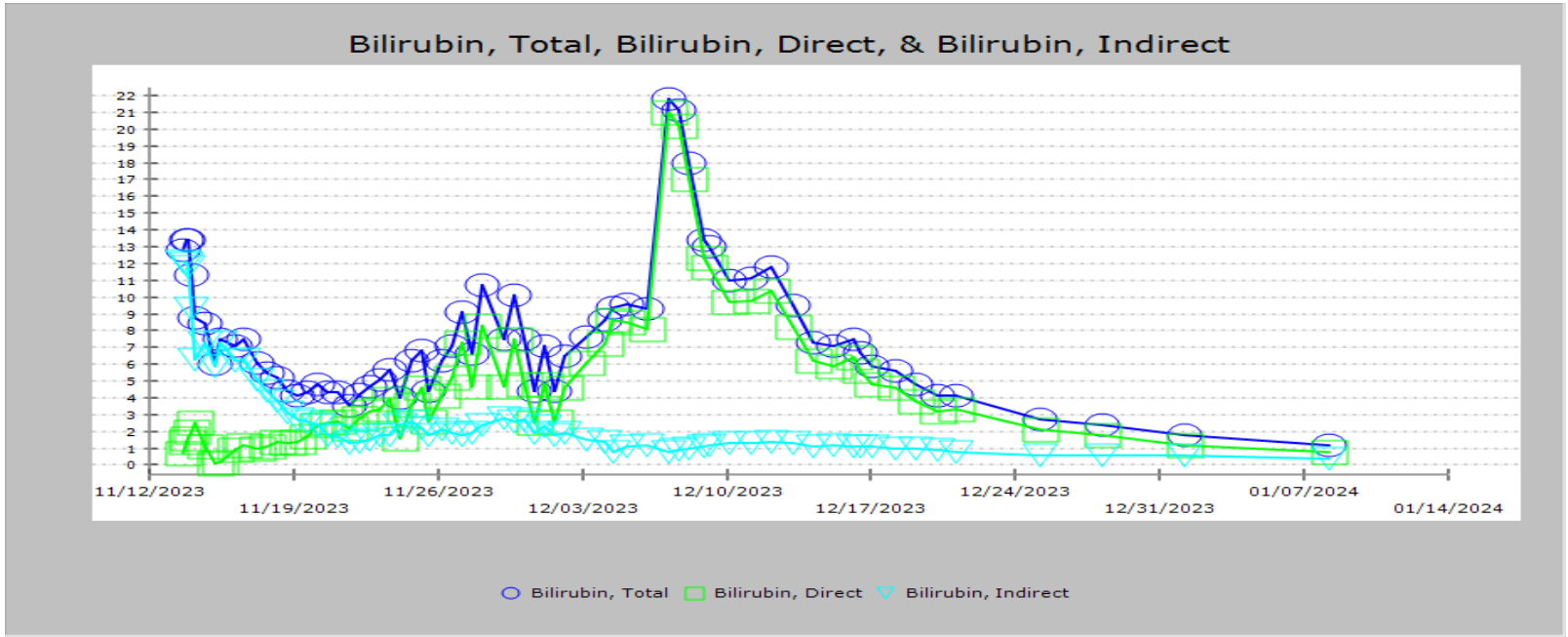
Direct bilirubin level: 20

Drop in hemoglobin to 6.4

CRRT, trial off for 24 hours

CRRT continued

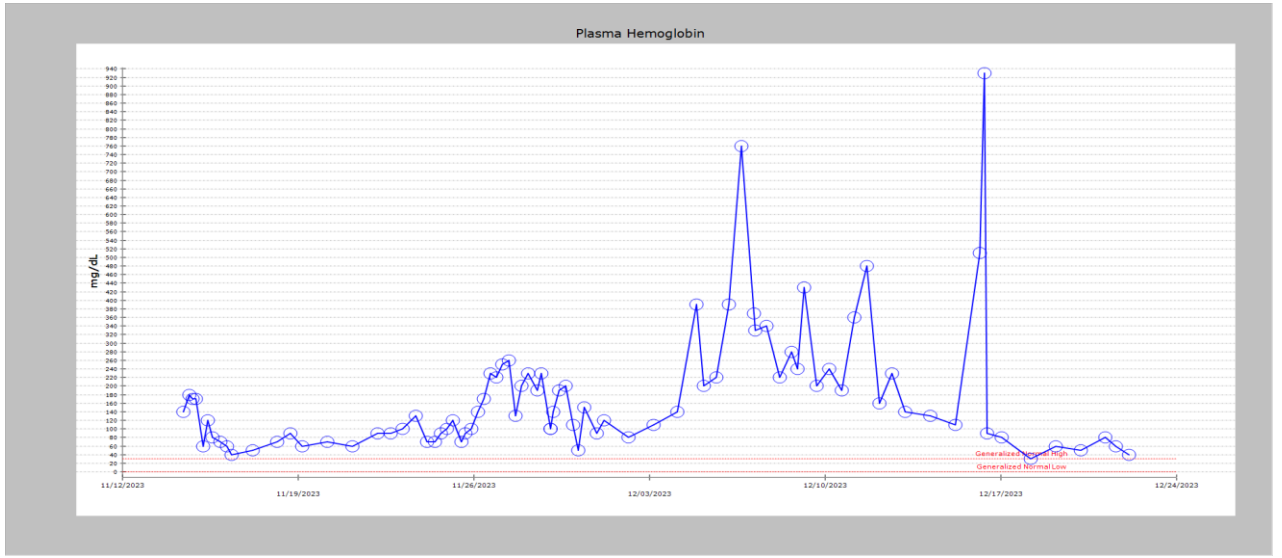
and stayed on-5 weeks total



Day 33

Brisk arterial bleeding from the right radial line was noted.

The line was removed.



Another month past by ……..

The patient is 2- months- old.

He is ready to go HOME!

Blood Bank Testing

	Blood type	ANTIBODY SCREEN	DAT: IgG GEL	OTHER
DAY 1	Infant: O + Mother: A+	0/3/2	4+	Sample sent to the reference lab. Confirmed: Anti E and c
DAY 3		0/2/1	1+	
DAY 7		0/2/1	1+	
DAY 10		0/2/1	0	Titer: anti E< 2 Titer: anti c < 2
DAY 17		0/2/1	0	
DAY 25		0/1/0	0	

Transfusions

Days	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31	32	33	34	35	36	TOTAL
RBC	4	2	4	3	3	1	2	3	1	0	0	1	0	0		2	2	1		1	1		1		2	1			1	1		1			1		39
PLT	2	2	3	2	2	2	2	2	2	1	2	4	2	3	4	1	2	2	1	2		1		1		1					1					1	48

Conclusion

- HDFN is a preventable disease.
- HDFN is a rare but potentially severe disease.
- Severe disease most commonly occurs due to alloantibodies directed against RhD, Rhc, K antigens.
- Non-Rh and non-K alloantibodies can cause mild form of the disease.

Thank you!

- Questions!
- Comments!

- Science without heart is ugly and pitiless.