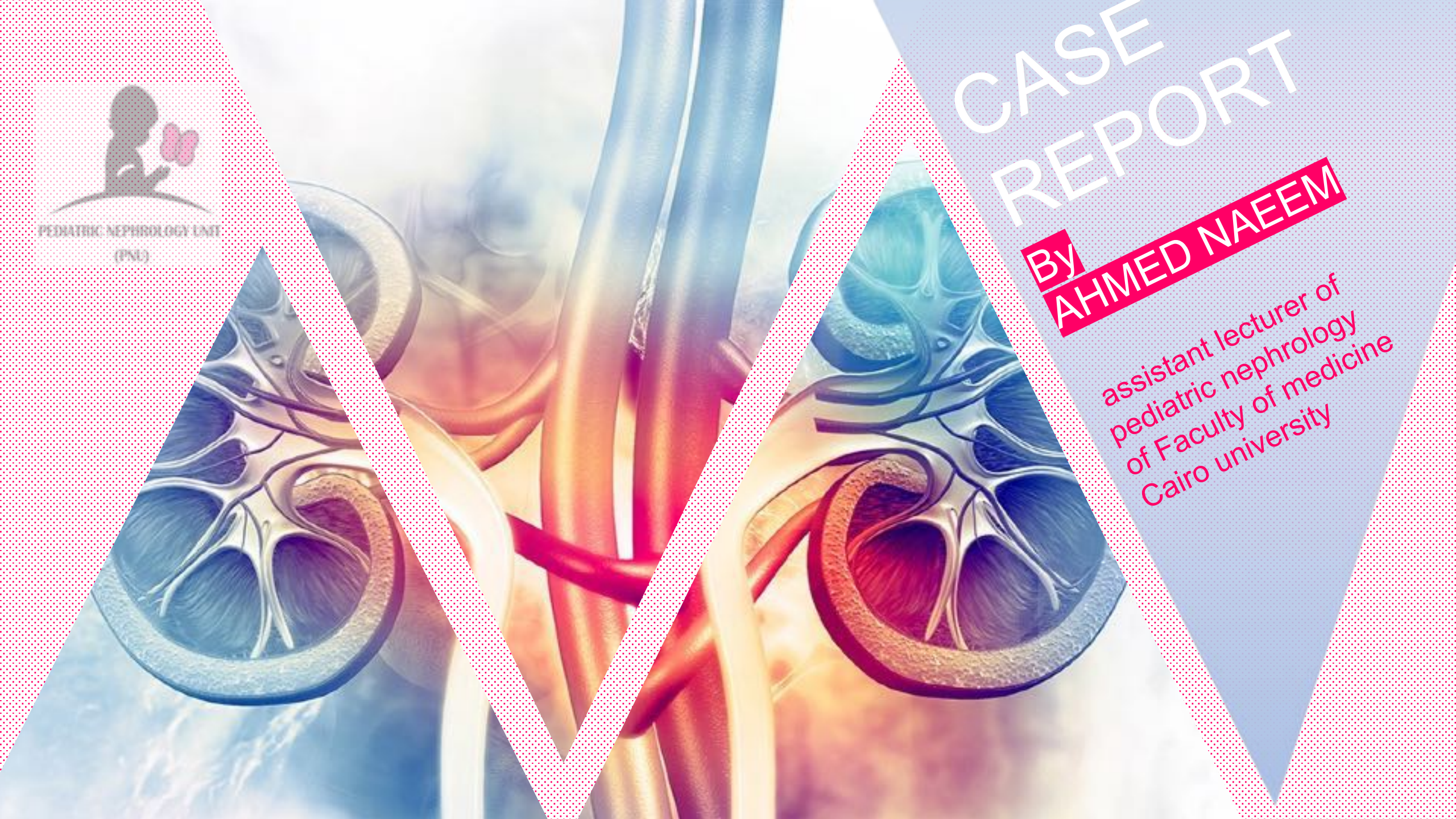
PEDIATRIC NEPHROLOGY UNIT

(PNU)



PEDIATRIC NEPHROLOGY UNIT
(PNU)

CASE REPORT

By
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of Faculty of medicine
Cairo university

MANAL MONTASER

11 Yr old

Consanguineous

4th sib

4 ♀

healthy sibs

previously

healthy

Condition started

08/2018

- with vomiting/diarrhea (bloody once)
- She received iv fluids & didn't improve
- She had normocytic normochromic anemia, received bl, then she was referred to us

Condition started

08/2018

- She had anemia, thrombocytopenia & impaired kidney functions
- she started PD then HD due to uremia and overload
- She was diagnosed clinically as HUS but didn't improve so she started plasmapheresis.

- 21 sessions of plasmapheresis were done and the patient improved (hematologically and renally)
- She was discharged with normal CBC

Creatinine 0.8 to follow up at the nephrology clinic

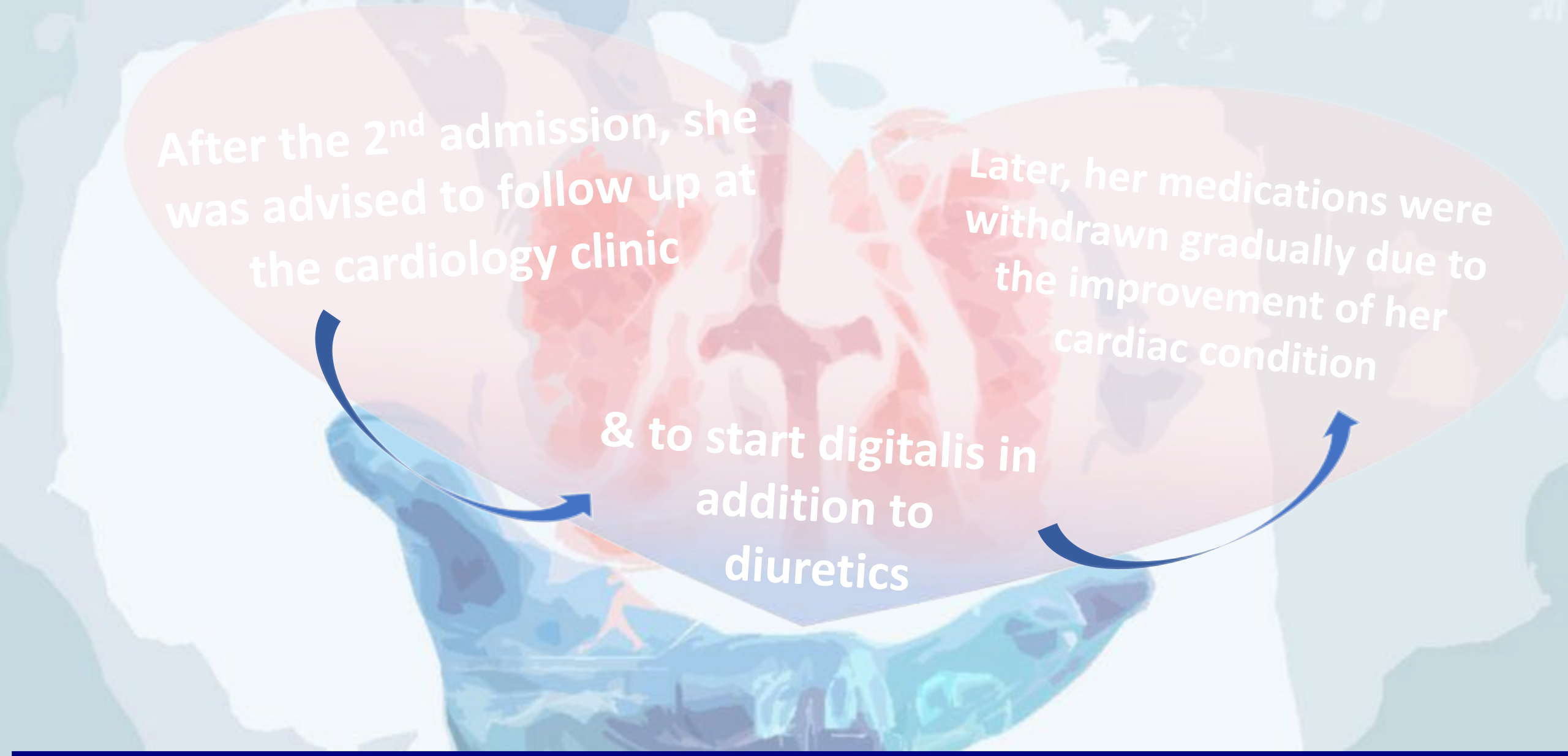
2 Months Later

She developed respiratory distress Due to HF

& was admitted at the ICU & put on MV& inotropes & diuretics

she was discharged after 1.5 months on anti HF medications


1 week after discharge
She was readmitted in the ICU with the same condition (cardiomyopathy) for 2 weeks



They suspected that her cardiomyopathy was 2r

TMA

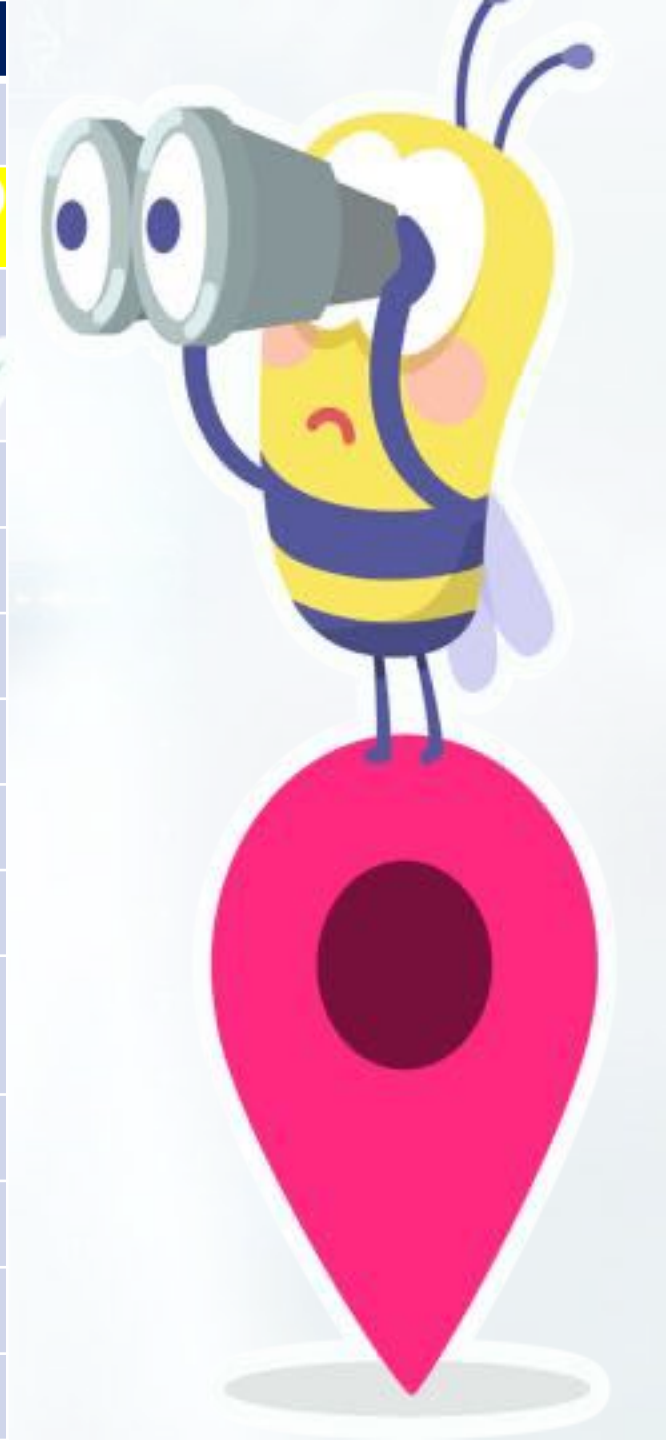
For 3.5 years after the first attack, she was on ACE inhibitors & was following at the CKD clinic, her CBC was

On May 2023, she presented to ER with severe pallor, dark urine & normal urine output, her weight & height were below the 3rd percentile despite being previously normal..

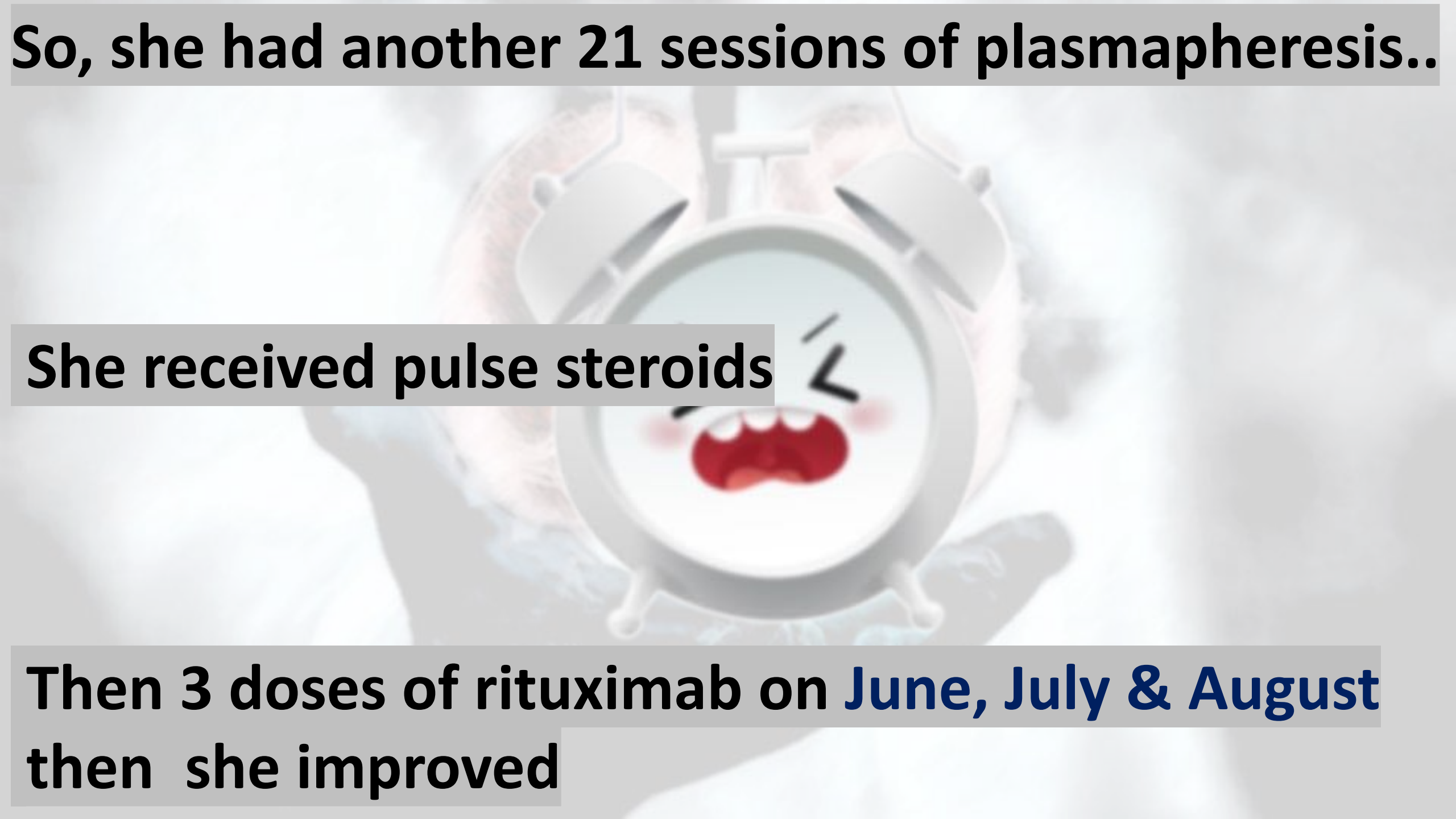
Her investigations showed normocytic normochromic anemia, thrombocytopenia & impaired kidney functions again..

RTX, Coombs, haptoglobin, C3 C4, fragmented RBCS were done
Antifactor H was +ve

Date	18/5	20/5	22/5
Blood Urea	112.3	140	
Serum Creatinine	1.64	2.4	
Hb	5.5	6.6	9
PLT	93	147	23
TLC	8.76	10.4	12.5
Reticulocyte Count	13.6 (High)		
Direct Coombs Test	-ve		
Fragmented RBCs	5.3		
Haptoglobin	0.1 (Low)		
Factor H		688 (300-800)	
Anti Factor H		Positive 97.1	
C3		98	
C4		23	
ANA with titre		-ve	



So, she had another 21 sessions of plasmapheresis..



She received pulse steroids

Then 3 doses of rituximab on June, July & August then she improved

INVESTIGATIONS

Date	22/2	18/5	20/5	22/5	1/6	10/6	11/6	26/6	1/7	9/7	5/8	5/9	25/10
Blood Urea	28	112.3	140		50		132		56	51			38
Serum Creatinine	0.4	1.64	2.4		1.17		1.6		0.95	0.8			0.5
Hb	13.5	5.5	6.6	9	9.6	5.1	8		11.2	10	9.1	11.6	10.9
PLT	289	93	147	23	173	55	54		119	156	214	269	231
TLC	5.5	8.76	10.4	12.5	12	13	21		2.46	5	6.4	6.2	5
A/C Ratio		7738						4737					
Reticulocyte Count		13.6 (High)											
Direct Coombs Test		-ve											
Fragmented RBCs		5.3											
Haptoglobin		0.1 (Low)						0.2 (0.3-2)					

Date	20/05	4/06
Factor H	688 (300-800)	
CRP	Negative	
ESR	145	
Anti Factor H	Positive 97.1	
Ferritin		2332.5
D-dimer		<0.5
Cardiac enzymes		normal
C3	98	90 (90-180)
C4	23	19.8 (10-40)
ANA with titre		Negative
ANCA		
Blood culture	No growth	

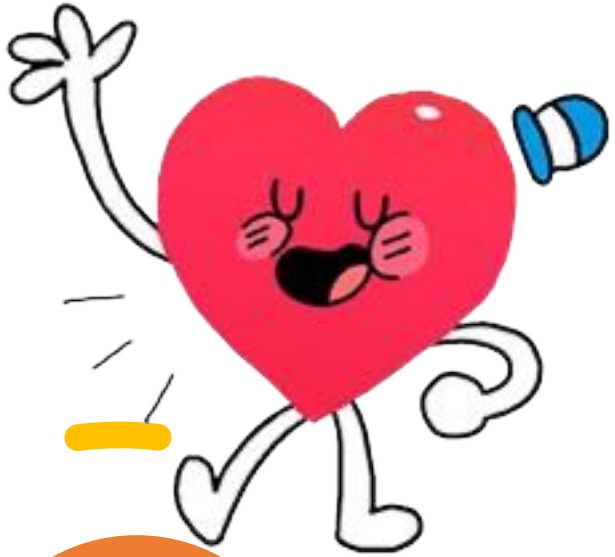
Date	Lab	Result
25/06/23	**Urine Analysis**	Pus cells 5-6, RBC 6-8 hemoglobin+, granular casts, pus cells casts
Date	**imaging**	**Result**
6/03/23	**PAUS**	Normal study
22/05/23	**Echocardiography**	Trivial MR, mild LV dilatation and hypertrophy, no vegetations, no pericardial effusion
19/06/23		Mild LV hypertrophy, mildly dilated RCA, for small dose aspirin if platelet count and coagulation profile permit
25/06/23		Better LV systolic function, mild RCA dilation for follow-up
16/07/23		Concentric LV hypertrophy with good systolic function, control blood pressure, normal coronaries

TO SUM UP

Our patient had anti factor H positive HUS; she recently went through relapse & two ICU admissions as she needed inotropes due to cardiomyopathy

Overall, she had 42 sessions of plasmapheresis, pulse steroid therapy& Rituximab

Thankfully, the patient is currently following at our clinic with normal kidney function tests, her last echo has good systolic function, and she stopped her anti HF medications



Thank You