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Sjögren Syndrome–Associated Immune Thrombocytopenia With Severe Postpartum Deterioration: A Case Report

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Case Report

Keywords: Sjögren syndrome, immune thrombocytopenia, postpartum period, case report

Posted Date: November 18th, 2026

DOI: https://doi.org/10.21203/rs.3.rs-11533314/v1

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Additional Declarations: No competing interests reported.

Abstract

Background

Sjögren syndrome (SS) may be complicated by secondary immune thrombocytopenia (ITP), yet detailed reports of its postpartum course are limited. We describe severe postpartum deterioration in a woman with SS-associated ITP, emphasizing continuity of care from discharge through long-term follow-up.

Case presentation:

A 34-year-old woman developed thrombocytopenia during early pregnancy and underwent cesarean delivery at 36 weeks and 6 days. Her platelet count recovered to 158 × 10^9/L by discharge on postpartum day 8 during treatment with prednisone, hydroxychloroquine, and cyclosporine. Feeling well, she stopped all three medications on day 16. On day 23, she was readmitted with a platelet count of 1 × 10^9/L, increased lochia, and petechiae. The patient was diagnosed with SS and concomitant primary biliary cholangitis. Combined rescue treatment included glucocorticoids, intravenous immunoglobulin, platelet transfusions, tacrolimus, and short-course recombinant human thrombopoietin. The platelet count reached 157 × 10^9/L on day 45 but fluctuated during subsequent treatment, measuring 106 × 10^9/L at approximately ten months postpartum.

Conclusions

This longitudinal account traces improvement before discharge, patient-initiated treatment discontinuation, readmission, and continuing outpatient care. This case highlights the need for an explicit medication plan, timely platelet reassessment, and coordinated obstetric, hematologic, and rheumatologic follow-up after childbirth in women with SS-associated ITP.

Background

Sjögren syndrome (SS) is a systemic autoimmune disease with glandular and extraglandular manifestations [1]. With the trend toward later childbearing and advances in assisted reproductive technologies, the number of pregnancies among women with primary SS may increase [2]. Secondary immune thrombocytopenia (ITP) is a recognized hematologic complication of SS. In a retrospective hospital-based cohort, ITP was identified in 35 of 291 patients with primary SS [3]. In women with SS-associated ITP, pregnancy and the postpartum period require coordinated obstetric, hematologic, and rheumatologic care.

Studies of pregnancy in SS have examined maternal and neonatal outcomes, and individual reports have described complex extraglandular manifestations during pregnancy [4, 5]. However, detailed descriptions of the postpartum course of SS-associated ITP remain scarce. A previous report documented

postpartum gingival and vaginal bleeding with profound thrombocytopenia, but did not describe a continuous antenatal-to-postpartum management trajectory [6].

Postpartum care for women with ITP includes reassessment of bleeding, platelet counts, and treatment needs, even when platelet counts have improved before discharge [7, 8]. We report a woman with SS-associated ITP whose course comprised initial postpartum improvement, patient-initiated treatment discontinuation, severe deterioration requiring readmission, and quantified follow-up to approximately ten months. The case illustrates how discharge planning and continuing outpatient care relate to the long-term course of a systemic autoimmune disease.

The CARE checklist is provided in Additional file 1.

Case presentation

Antenatal course

A 34-year-old woman (gravida 3, para 1) had a history of cesarean delivery. A platelet count of 135 × 10^9/L had been documented in the preceding year. In early pregnancy, laboratory testing showed a platelet count of 43 × 10^9/L and a hemoglobin concentration of 119 g/L, with unremarkable red-cell morphology. Autoimmune testing at that time revealed antinuclear antibody positivity at a titer of 1:320, along with positive anti-Sjögren syndrome type A antibody (anti-SSA) and anti-Ro52 (52-kDa Ro antigen) antibodies; anti-Sjögren syndrome type B antibody (anti-SSB), anti-double-stranded DNA, and anti-Smith (anti-Sm) antibodies were negative. Further history revealed approximately six months of dry mouth and five months of dry eyes. SS was suspected, and hydroxychloroquine 200 mg daily was prescribed.

When her platelet count subsequently decreased to 28 × 10^9/L at 31 weeks and 2 days, prednisone acetate 10 mg daily and cyclosporine 50 mg daily were prescribed. At 34 weeks and 2 days, the platelet count was 16 × 10^9/L, hemoglobin 119 g/L, alanine aminotransferase and aspartate aminotransferase both 23 U/L, lactate dehydrogenase 185 U/L, and creatinine 43 μmol/L. Given the risk of preterm delivery, dexamethasone was administered to improve neonatal outcomes. Two days before delivery, an outpatient platelet count was 56 × 10^9/L; prednisone and cyclosporine were reduced on medical advice to 5 mg and 25 mg daily, respectively.

Delivery and initial postpartum improvement

At 36 weeks and 6 days, she presented with preterm premature rupture of the membranes and a platelet count of 45 × 10^9/L. The count increased to 81 × 10^9/L after preoperative platelet transfusion. Cesarean delivery resulted in a male infant weighing 2320 g, with Apgar scores of 9 and 10. Uterine atony and needle-site oozing were documented, with estimated blood loss of 800 mL. Management included uterotonics, tranexamic acid, uterine tamponade and compression measures, and additional platelet and plasma transfusions. The day of delivery is designated postpartum day 0.

Postpartum hypertension, with a maximum recorded blood pressure of 153/105 mmHg, was treated with labetalol. The platelet count decreased to 30×10^9/L on day 4, prompting another platelet transfusion. Treatment comprised prednisone 15 mg daily, hydroxychloroquine 200 mg twice daily, and cyclosporine 50 mg in the morning and 25 mg in the evening. By discharge on day 8, the platelet count had increased to 158×10^9/L and hemoglobin was 94 g/L. She was instructed to continue the same three-drug regimen after discharge.

Treatment discontinuation and readmission

Feeling well, the patient stopped prednisone, hydroxychloroquine, and cyclosporine on day 16 without medical advice. On postpartum day 23, she returned to the outpatient clinic with a 3-day history of increased lochia and intermittent gingival bleeding. Laboratory testing revealed a platelet count of 1×10^9/L and a hemoglobin concentration of 120 g/L. Physical examination revealed cutaneous petechiae. No platelet measurement was available between discontinuation and readmission. The previous regimen was restarted, and platelets were transfused.

Further investigations showed positive platelet-specific antibodies, an immunoglobulin G concentration of 19.71 g/L, creatinine of 49 μmol/L, and unremarkable red-cell morphology. Alanine aminotransferase was 91 U/L and aspartate aminotransferase 77 U/L. Bone marrow examination showed increased megakaryocytes with impaired platelet production, with no abnormal cell population detected by flow cytometry. Salivary gland scintigraphy demonstrated moderate functional impairment of both submandibular glands and mild functional impairment of the parotid glands. Based on ocular and oral symptoms, anti-SSA antibody positivity, and objective evidence of salivary gland involvement, a diagnosis of SS-associated ITP was established following multidisciplinary evaluation.

Antimitochondrial M2 antibody testing was strongly positive, with a previously recorded level exceeding 400 RU/mL. Serum alkaline phosphatase was also elevated, whereas hepatobiliary ultrasonography was unremarkable. Concomitant primary biliary cholangitis (PBC) was diagnosed, and ursodeoxycholic acid was included in her continuing treatment [9]. The maternal autoantibody profile and related test results are summarized in Table 1.

Table 1
Maternal autoantibody profile and related testing

Antibody or test	Result
ANA and related autoantibodies	
ANA	1:80, nuclear and cytoplasmic speckled
Anti-SSA	+++
Anti-Ro52	+++
Anti-SSB	Negative
Anti-dsDNA	Negative
Anti-Sm	Negative
Additional ANA-profile antibodies[a]	Negative
Autoimmune liver serology	
AMA-M2	> 400 RU/mL
AMA (reported separately)	Negative
Other autoimmune liver antibodies[b]	< 2.00 RU/mL each
Antiphospholipid testing	
aCL (IgG, IgM and IgA)	Within reference range[c]
Anti-β2GPI (IgG, IgM and IgA)	Within reference range[c]
LA1/LA2 ratio	1.34 (reference 0.8–1.2)[d]
Additional serologic testing	
Platelet-specific antibodies	Positive
Rheumatoid factor	36.60 IU/mL (reference < 20 IU/mL)

Antibody or test	Result
ANA and related autoantibodies	
+++ reproduces the laboratory's qualitative grading.	
[a] Anti-nRNP, anti-ribosomal P, anti-nucleosome, anti-histone, anti-PCNA, anti-Jo-1, anti-PM-Scl, anti-Scl-70 and anticentromere antibodies.	
[b] Anti-LKM-1, anti-LC-1, anti-SLA/LP, anti-smooth muscle, anti-sp100 IgG and anti-gp210 IgG; upper reference limit, 20 RU/mL. AMA and AMA-M2 were reported as separate tests.	
[c] All three isotypes were below 20 RU/mL. [d] The measured LA1/LA2 ratio is shown; a qualitative LA interpretation was not provided.	
ANA, antinuclear antibody; anti-SSA, anti-Sjögren syndrome type A antibody; anti-SSB, anti-Sjögren syndrome type B antibody; Ro52, 52-kDa Ro antigen; dsDNA, double-stranded DNA; Sm, Smith antigen; AMA, antimitochondrial antibody; M2, mitochondrial antigen type 2; aCL, anticardiolipin antibody; β2GPI, β2-glycoprotein I; LA, lupus anticoagulant; IgG, IgM, and IgA, immunoglobulin G, M, and A.	
nRNP, nuclear ribonucleoprotein; PCNA, proliferating cell nuclear antigen; Jo-1, histidyl-transfer RNA synthetase; PM-Scl, polymyositis–scleroderma antigen; Scl-70, DNA topoisomerase I; LKM-1, liver–kidney microsomal antigen type 1; LC-1, liver cytosol antigen type 1; SLA/LP, soluble liver antigen/liver–pancreas antigen; sp100, nuclear protein sp100; gp210, nuclear pore glycoprotein 210; RU, relative units; IU, international units.	

[Insert Table 1 here]

Intravenous methylprednisolone 500 mg daily was administered on days 25–27, together with a single 10 g dose of intravenous immunoglobulin on day 25. Hydroxychloroquine and cyclosporine were initially continued, and additional platelets were transfused. The platelet count remained at 1×10^9/L on day 26, increased to 10×10^9/L on day 27, and was 8×10^9/L on day 28. On day 28, cyclosporine was replaced by tacrolimus 1 mg twice daily, recombinant human thrombopoietin was started at 15,000 units subcutaneously once daily, and methylprednisolone was reduced to 120 mg daily. The platelet count reached 38×10^9/L on day 31, when recombinant human thrombopoietin was discontinued, and 91×10^9/L on day 35. Bleeding manifestations gradually subsided during combined treatment.

Long-term maternal and infant follow-up

On day 37, she was discharged on oral methylprednisolone 60 mg daily, with a planned reduction of 4 mg each week, tacrolimus 1 mg twice daily, hydroxychloroquine 200 mg twice daily, and ursodeoxycholic acid 250 mg twice daily. The latest platelet measurement before discharge was 91×10^9/L on day 35. The platelet count reached 157×10^9/L on day 45 and 224×10^9/L on day 66. On day 94, the count was 77×10^9/L while the recorded treatment regimen included methylprednisolone 12 mg daily and oral tacrolimus. At approximately ten months postpartum, the platelet count was 106×10^9/L; prednisone

was increased from 5 to 10 mg daily, with hydroxychloroquine prescribed. Platelet counts continued to fluctuate during follow-up despite ongoing treatment. Table 2 summarizes the main changes in treatment and follow-up, and Fig. 1 shows the recorded platelet counts.

Table 2
Key transitions in postpartum treatment and follow-up

Time	Platelet count (× 10^9/L)	Clinical course and treatment
Early pregnancy	43	Thrombocytopenia identified; sicca symptoms and anti-SSA and anti-Ro52 positivity prompted evaluation for SS.
34 weeks 2 days	16	Persistent antenatal thrombocytopenia despite treatment.
Day 0	45 before transfusion; 81 after transfusion	Cesarean delivery at 36 weeks and 6 days; uterine atony and operative oozing; estimated blood loss 800 mL.
Day 8	158	Discharged on prednisone 15 mg/day, HCQ 200 mg twice daily, and cyclosporine 50 mg in the morning and 25 mg in the evening.
Day 16	Not measured	Patient stopped all three medications because she felt well.
Day 23	1	Readmitted with increased lochia and petechiae; previous medications restarted and platelets transfused.
Days 25–35	Day 26: 1; day 28: 8; day 31: 38; day 35: 91	Pulse-dose MP and IVIG; cyclosporine changed to tacrolimus; short-course rhTPO. Bleeding subsided.
Day 37	Latest result: 91 on day 35	Discharged on oral MP, tacrolimus, HCQ, and UDCA with a planned MP taper.
Days 45, 66, and 94	157, 224 and 77	Recovery followed by fluctuation during ongoing treatment.
Day 311	106	Prednisone increased from 5 to 10 mg/day; HCQ and UDCA prescribed.

Day 0 denotes the day of delivery. No platelet count was recorded at treatment discontinuation or on day 37. Anti-SSA, anti-Sjögren syndrome type A antibody; Ro52, 52-kDa Ro antigen; HCQ, hydroxychloroquine; IVIG, intravenous immunoglobulin; MP, methylprednisolone; rhTPO, recombinant human thrombopoietin; SS, Sjögren syndrome; UDCA, ursodeoxycholic acid.

[Insert Table 2 here]

The infant developed a rash, with positive tests for anti-SSA and anti-Ro52 antibodies. The rash subsequently resolved. No thrombocytopenia or atrioventricular block was documented. Neonatal lupus was considered but remained unconfirmed.

Discussion and Conclusions

Postpartum deterioration after improvement at discharge

Maintaining disease control after discharge was the main challenge in this woman with SS-associated ITP, despite her initial postpartum recovery. At discharge, her platelet count was 158 × 10^9/L while she was receiving prednisone, hydroxychloroquine, and cyclosporine. She stopped all three medications after feeling well and was readmitted one week later with a platelet count of 1 × 10^9/L and bleeding. This sequence highlights the clinical importance of the transition from obstetric discharge to outpatient management.

Reviews of ITP in pregnancy emphasize individualized postpartum treatment and continued assessment of bleeding and platelet counts [7, 8]. For women with SS-associated ITP, this care also needs to address the underlying autoimmune disease after obstetric discharge. A practical discharge plan should specify the medication regimen, the timing of platelet reassessment, and the clinician responsible for dose changes. Counseling should explain why treatment may need to continue even when patients feel well or their platelet counts have improved, and should identify increased lochia, new petechiae, and mucosal bleeding as reasons for prompt evaluation.

The bleeding episodes required assessment of both obstetric and hematologic factors. Uterine atony and operative oozing occurred at delivery, whereas increased lochia and mucocutaneous bleeding accompanied profound thrombocytopenia at readmission. These findings support continued collaboration between obstetric and hematology teams after the delivery admission.

A rare presentation with added longitudinal detail

Our targeted literature review identified one previous individually described case of postpartum SS-associated ITP with vaginal bleeding: the 22-year-old patient reported by Naeem et al [6]. However, that report did not detail the antenatal platelet trajectory, specify the exact postpartum day of presentation, or clarify whether treatment had been interrupted before presentation. Follow-up was qualitative, with neither serial platelet counts nor a clearly stated duration. These omissions make the sequence of events and subsequent course difficult to reconstruct.

This case documents a rarely reported presentation of SS-associated ITP with severe postpartum deterioration and provides a detailed longitudinal account. Antenatal platelet measurements show the course before delivery. The postpartum record gives the exact timing of improvement at discharge, discontinuation of three medications, and readmission, followed by serial counts documenting fluctuations through approximately ten months. Objective evidence of glandular involvement further documents the underlying SS. The record thus describes both a rare postpartum presentation and the challenges of continuing maternal care.

Of 81 pregnant women with ITP in one cohort, two had SS, but their individual postpartum courses were not reported [10]. In nonpregnant patients with SS-associated ITP, Xu et al reported recurrence during treatment withdrawal or dose reduction [11]. By contrast, Weinstein Mezan et al described severe postpartum worsening despite ongoing treatment in primary ITP [12]. These reports describe different patterns of disease control. SS-associated ITP should still be distinguished from primary ITP and other causes of thrombocytopenia.

Coordinated treatment and follow-up beyond delivery

The established diagnosis of SS provides the clinical context for this patient's secondary ITP. The early onset, severity, and persistence of thrombocytopenia after delivery supported an immune process rather than uncomplicated gestational thrombocytopenia [7]. Coexisting PBC was confirmed according to the 2018 guidance from the American Association for the Study of Liver Diseases. SS and PBC are recognized coexisting autoimmune conditions [9]. Their coexistence requires coordinated rheumatologic and hepatologic care alongsideas well as platelet monitoring.

During follow-up, the platelet count increased from 157 to 224 × 10^9/L, then decreased to 77 × 10^9/L, and subsequently rose to 106 × 10^9/L. These fluctuations occurred during ongoing treatment, highlighting the importance of continued platelet monitoring after discharge. For women with SS-associated ITP, care after childbirth therefore needs to cover acute bleeding, treatment review and adjustment, and follow-up of the underlying systemic autoimmune disease.

In a single case, we cannot determine whether stopping treatment caused the deterioration or assess the contribution of each component of rescue treatment. The intervals between platelet measurements also prevent us from identifying when the decline began.

Conclusions

SS-associated ITP can require ongoing management after an initially favorable postpartum recovery. After improving before discharge, this patient stopped treatment on her own, was readmitted with severe deterioration, and had further platelet fluctuations during approximately ten months of follow-up. The case highlights the clinical value of coupling improvement at discharge with a clear medication plan, timely reassessment, and coordinated care across obstetrics, hematology, and rheumatology.

Abbreviations

aCL
anticardiolipin antibody
AMA
antimitochondrial antibody
AMA-M2

antimitochondrial M2 antibody
ANA
antinuclear antibody
β2GPI
β2-glycoprotein I
dsDNA
double-stranded DNA
gp210
nuclear pore glycoprotein 210
HCQ
hydroxychloroquine
IgA
immunoglobulin A
IgG
immunoglobulin G
IgM
immunoglobulin M
ITP
immune thrombocytopenia
IU
international units
IVIG
intravenous immunoglobulin
Jo-1
histidyl-transfer RNA synthetase
LA
lupus anticoagulant
LC-1
liver cytosol antigen type 1
LKM-1
liver–kidney microsomal antigen type 1
M2
mitochondrial antigen type 2
MP
methylprednisolone
nRNP
nuclear ribonucleoprotein
PBC
primary biliary cholangitis
PCNA

proliferating cell nuclear antigen
PM-Scl
polymyositis–scleroderma antigen
rhTPO
recombinant human thrombopoietin
Ro52
52-kDa Ro antigen
RU
relative units
Scl-70
DNA topoisomerase I
SLA/LP
soluble liver antigen/liver–pancreas antigen
Sm
Smith antigen
sp100
nuclear protein sp100
SS
Sjögren syndrome
anti-SSA
anti-Sjögren syndrome type A antibody
anti-SSB
anti-Sjögren syndrome type B antibody
UDCA
ursodeoxycholic acid.

Declarations

Ethics approval and consent to participate

This case report was approved by the ethics committee of the Fourth Hospital of Zhejiang University, China, and adhered to the principles of the Declaration of Helsinki.

Consent for publication

The requirement for patient consent for publication was waived based on the protocol of the retrospective case report approved by the ethics committee of the Fourth Hospital of Zhejiang University, China.

Competing interests

The authors declare no competing interests.

Funding

Not applicable.

Author Contribution

P.Y.C. wrote the main manuscript text and prepared figure 1. C.X.Y. read and approved the final version of the manuscript.

Acknowledgement

We would like to express our deep gratitude to the patient and their family members for agreeing to participate in this study.

Data Availability

All data supporting the findings of this study are available within the paper.

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Figures

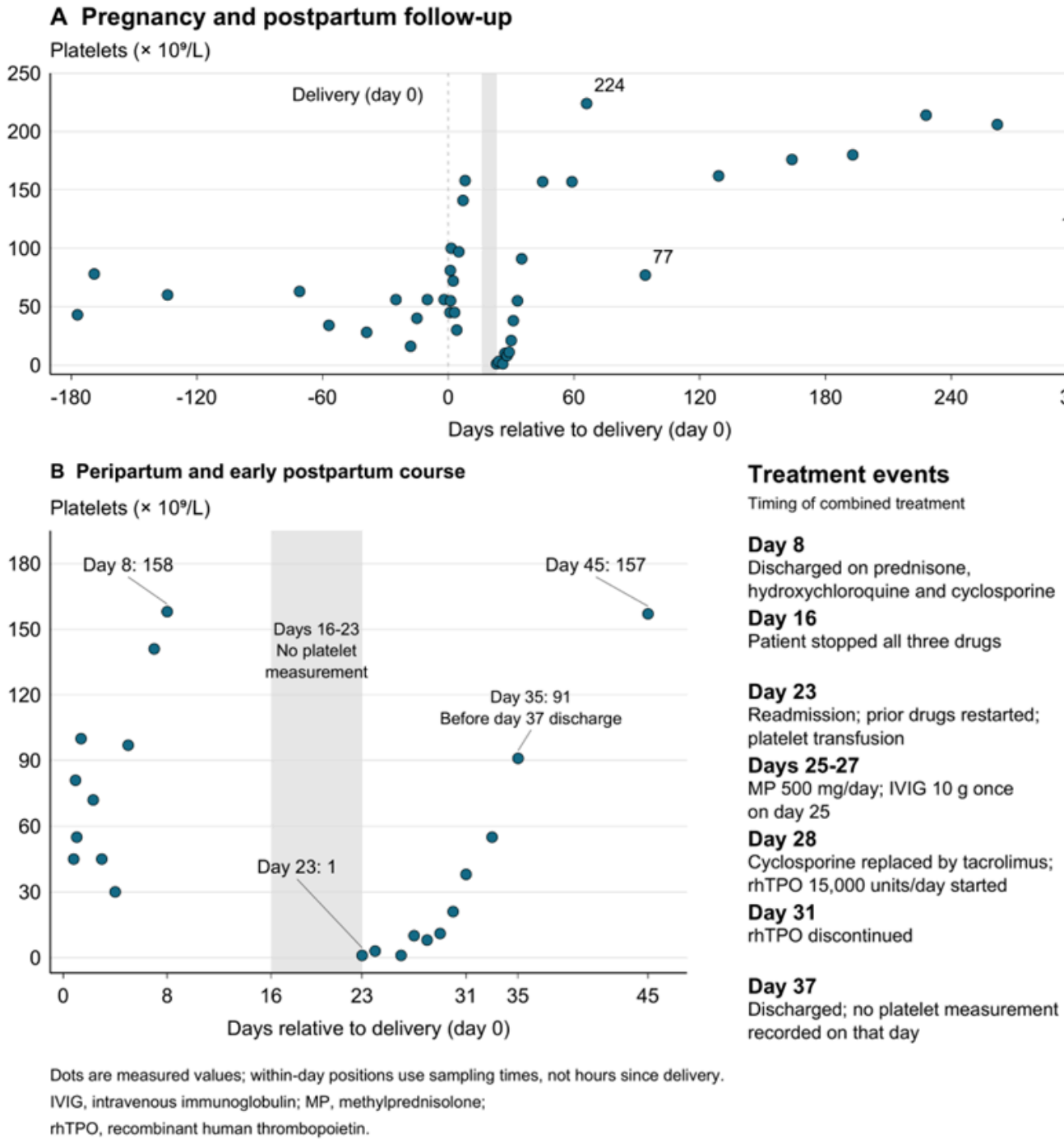


Figure 1

Platelet measurements across pregnancy and postpartum care.

Panel A shows the recorded pregnancy and follow-up measurements; panel B enlarges the peripartum and early postpartum course. Day 0 denotes the day of delivery. Dots are measured values without interpolation. The shaded interval between days 16 and 23 marks the period from treatment discontinuation to readmission, during which no platelet measurement was recorded. Within-day positions use recorded sampling times where available, not elapsed hours from birth. The platelet count of 91 × 10^9/L was obtained on day 35, before discharge on day 37. Treatment annotations show their

timing rather than separate drug effects. IVIG, intravenous immunoglobulin; MP, methylprednisolone; rhTPO, recombinant human thrombopoietin.