European Journal of Clinical Pharmacy

Print ISSN: 2385-409X **Online ISSN**: 3105-0409
Website: https://farmclin.com/

Case Report

Obstetric Implications of Maternal Sjögren's Syndrome: From Fetal Complete Heart Block to a Successful Subsequent Pregnancy

Name of Author:
PANKTI PARIKH[1] SHRIDEVI METGUD[2] PRAVIN JAIN[3] ANITA DALAL[4]

Affiliation: *[1]Postgraduate, Dept of OBGY, Jawaharlal Nehru Medical College and KLES Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi*
[2]Associate Professor, Dept of OBGY, Jawaharlal Nehru Medical College and KLES Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi
[3]Consultant, Dept of Rheumatology, Jawaharlal Nehru Medical College and KLES Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi
[4]Professor, Dept of OBGY, Jawaharlal Nehru Medical College and KLES Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi

Corresponding Author:
Dr. Pankti Parikh

Received: 31-11-2025
Revised: 20-12-2025
Accepted: 30-12-2025
Published: 05-02-2026

DOI: 10.61336/ejcp/25-12-584

Abstract: **Background:** Primary Sjögren's syndrome (pSS) is a chronic systemic autoimmune disease that predominantly affects women in the third to fifth decades of life. Diagnosis in adolescence or early adulthood is uncommon, making this case notable for the early onset of disease at 20 years of age. Hypokalemic paralysis is a particularly uncommon presentation of distal renal tubular acidosis (dRTA), a less common renal symptom that affects only 3–5% of patients with primary Sjögren's syndrome. In maternal Sjögren's syndrome, anti-Ro/SSA and anti-La/SSB antibodies greatly increase the likelihood of fetal cardiac involvement, specifically congenital heart block (CHB).

Case description: Fetal bradycardia with pericardial effusion was found when a 23-year-old woman with primary Sjögren's disease presented for a routine anomaly scan during her first pregnancy. Fetal echocardiography confirmed complete heart block, leading to pregnancy termination. In her subsequent pregnancy, early initiation of hydroxychloroquine and intensive fetal cardiac surveillance resulted in a normal fetal cardiac course. Delivery at 34 weeks was indicated for late-onset fetal growth restriction (FGR) with abnormal Doppler findings, and a healthy female neonate was delivered with a normal postnatal echocardiogram.

Conclusion: Early detection, hydroxychloroquine therapy, and structured fetal surveillance are essential for optimizing outcomes and are pivotal in anti-SSA–positive pregnancies. Although the first pregnancy was complicated by irreversible fetal complete heart block, appropriate antenatal surveillance facilitated a successful outcome in the next gestation.

Clinical significance: The case highlights the early onset of the autoimmune disease in the absence of classical sicca symptoms. Proper preconception counselling, adherence to medical therapy and adequate fetal surveillance are the key to a successful pregnancy outcome.

Keywords: Sjogren's syndrome, pregnancy, fetal heart block, hydroxychloroquine

INTRODUCTION

Primary Sjögren's syndrome (pSS) is a chronic systemic autoimmune disorder characterized by lymphocytic infiltration of exocrine glands, leading classically to xerophthalmia and xerostomia. It predominantly affects women, most commonly presenting in the third to fifth decades of life (1).

However, disease onset during adolescence or early adulthood is uncommon and often poses diagnostic challenges due to the absence of classical sicca symptoms and the predominance of extraglandular manifestations. Early-onset Sjögren's syndrome may therefore remain underdiagnosed until systemic complications or pregnancy-related issues bring the condition to clinical attention (2).

Pregnancy in women with pSS is associated with unique maternal and fetal risks, largely mediated by circulating autoantibodies, particularly anti-Ro/SSA and anti-La/SSB antibodies. These antibodies can cross the placenta beginning in the late first trimester and exert pathogenic effects on fetal tissues (3). Among the most serious fetal complications is congenital heart block (CHB), a manifestation of neonatal lupus syndrome, which results from immune-mediated injury to the fetal atrioventricular conduction system. CHB typically develops between 16 and 24 weeks of gestation and may progress rapidly from first-degree block to irreversible complete heart block (4).

Although the absolute risk of CHB in a first anti-SSA–positive pregnancy is relatively low, estimated at approximately 2–5%, the recurrence risk increases substantially to 15–18% following a previously affected pregnancy. Once complete heart block is established, reversal is exceedingly rare, often necessitating lifelong pacemaker placement and carrying significant risks of fetal hydrops, intrauterine demise, and neonatal morbidity (5). Consequently, early identification of high-risk pregnancies and implementation of preventive strategies are critical components of obstetric management in women with Sjögren's syndrome (6).

Hydroxychloroquine has emerged as a cornerstone therapy in anti-SSA–positive pregnancies. Beyond its established role in controlling maternal autoimmune disease activity, hydroxychloroquine has been shown to reduce interferon-mediated inflammation and decrease the recurrence risk of fetal cardiac manifestations by approximately 50% when initiated preconceptionally or early in pregnancy (7). Current rheumatology and obstetric guidelines support the continuation of hydroxychloroquine throughout pregnancy owing to its favorable safety profile for both mother and fetus (8).

In addition to fetal cardiac complications, pregnancies complicated by Sjögren's syndrome are associated with an increased incidence of placental dysfunction, fetal growth restriction, hypertensive disorders of pregnancy, preterm birth, and higher rates of cesarean delivery (9). These risks persist even in the absence of fetal cardiac involvement, underscoring the need for comprehensive antenatal surveillance extending beyond fetal echocardiography alone (10). Multidisciplinary care involving obstetricians, fetal medicine specialists, rheumatologists, and nephrologists is essential to optimize both maternal health and perinatal outcomes (11).

This case report highlights a rare presentation of early-onset primary Sjögren's syndrome with extraglandular renal involvement and illustrates the contrasting outcomes of two consecutive pregnancies—one complicated by irreversible fetal complete heart block and the subsequent pregnancy resulting in a favorable neonatal outcome following strict adherence to hydroxychloroquine therapy and structured fetal surveillance. The case underscores the critical importance of preconception counseling, medication compliance, and vigilant antenatal monitoring in improving outcomes in high-risk autoimmune pregnancies.

CASE PRESENTATION

Initial Presentation and Clinical Examination

A 20-year-old unmarried nulligravida with no known comorbidities presented to the emergency department with a gradual onset of bilateral lower limb weakness and difficulty in rising from bed. There was no associated sensory loss, bowel or bladder involvement, history of trauma, or preceding illness. She denied symptoms suggestive of electrolyte imbalance or neuromuscular disease, including diarrhoea, polyuria, dysphagia, or muscle cramps. Notably, there was no history of classical sicca symptoms such as dry eyes or dry mouth.

On clinical examination, neurological assessment revealed reduced motor power, graded as 3/5 in the right lower limb and 4/5 in the left lower limb. Cranial nerve examination was normal, and no sensory level deficits were identified. Systemic examination did not reveal any additional abnormalities.

Laboratory Evaluation and Etiological Workup

Initial laboratory investigations demonstrated severe hypokalemia, with a serum potassium level of 1.9 mEq/L, accompanied by metabolic acidosis. Serum bicarbonate was reduced to 16 mEq/L, and serum chloride was elevated at 113 mEq/L, while serum sodium was within normal limits at 142 mEq/L. Arterial blood gas analysis revealed a pH of 7.45 with a low bicarbonate level of 12 mEq/L, consistent with

compensated metabolic acidosis. Renal function was preserved, with a serum creatinine of 1.2 mg/dL. Hematological parameters showed hemoglobin of 10.4 g/dL, total leukocyte count of 12.6 $\times 10^3/\mu L$, and platelet count of 272 $\times 10^3/\mu L$. Thyroid function testing revealed mildly elevated thyroid-stimulating hormone levels at 7.55 mIU/L. Serum calcium levels were marginally reduced, with total calcium of 8.0 mg/dL and free calcium of 1.16 mg/dL.

Urine analysis showed an inappropriately high urine pH of 7.5 in the presence of metabolic acidosis, along with urinary potassium excretion of 14.8 mEq/L and urine creatinine of 21.06 mg/dL. The presence of a normal anion gap metabolic acidosis with a positive urine anion gap supported the diagnosis of distal renal tubular acidosis (Type 1).

Given the differential diagnosis of distal renal tubular acidosis, including familial, metabolic, granulomatous, and autoimmune etiologies, a detailed autoimmune workup was performed after exclusion of secondary causes. Antinuclear antibody profiling revealed strong positivity for anti-SSA, anti-SSB, and anti-Ro 52 antibodies. In the absence of other systemic autoimmune features and classical sicca manifestations, the patient was diagnosed with primary Sjögren's syndrome presenting with extraglandular renal involvement in the form of distal renal tubular acidosis and hypokalemic paralysis.

Initial Management

The patient was initiated on hydroxychloroquine 200 mg once daily along with potassium citrate supplementation for correction of hypokalemia and metabolic acidosis. With treatment, her electrolyte levels stabilized, and she remained under regular clinical follow-up.

First Pregnancy and Adverse Fetal Outcome

Three years later, in 2024, the patient presented at 19 weeks and 6 days of gestation during her first pregnancy for a routine anomaly scan. Ultrasonography revealed severe fetal bradycardia, with a fetal heart rate of approximately 70 beats per minute, along with pericardial effusion. Urgent fetal echocardiography confirmed complete (third-degree) atrioventricular heart block with features of fetal hydrops in the presence of a structurally normal heart. On further evaluation, it was noted that the patient had been non-compliant with hydroxychloroquine therapy during the preconceptional and antenatal period. The findings were consistent with immune-mediated injury to the fetal atrioventricular conduction system secondary to transplacental transfer of maternal anti-Ro antibodies.

The patient and her family received multidisciplinary counseling regarding the poor prognosis associated with early-onset complete heart block, including the high risk of fetal demise and the potential need for permanent pacemaker implantation in the neonatal period. Given the irreversible nature of the condition and unfavorable prognosis, the couple opted for termination of pregnancy. Post-termination counseling emphasized the increased recurrence risk of congenital heart block, estimated at approximately 18%, in subsequent pregnancies and highlighted the importance of strict adherence to hydroxychloroquine therapy.

Subsequent Pregnancy and Favorable Outcome

The patient conceived spontaneously eight months later. Hydroxychloroquine 200 mg once daily was continued throughout the preconceptional and antenatal period. Electrolyte levels and renal function were optimized, and low-dose aspirin was initiated. A detailed early anomaly scan at 16 weeks of gestation demonstrated normal fetal cardiac anatomy and rhythm.

In view of her anti-SSA and anti-SSB positivity and previous history of fetal complete heart block, intensive fetal surveillance with weekly sonographic and cardiac assessments was undertaken between 16 and 24 weeks of gestation. No conduction abnormalities were detected during this period.

At 34 weeks of gestation, the patient developed late-onset fetal growth restriction, with Doppler studies revealing increased umbilical artery resistance and a reduced cerebroplacental ratio, indicative of fetoplacental insufficiency. An emergency lower-segment cesarean section was performed based on maternal request and obstetric indications. A healthy female neonate weighing 2.2 kg was delivered, and postnatal echocardiography confirmed normal cardiac structure and rhythm.

DISCUSSION

A chronic systemic autoimmune disorder, primary Sjögren's syndrome (pSS) primarily affects women in their third to fifth decades of life. Diagnosis in adolescence or early adulthood is uncommon, making this case notable for the early onset of disease at 20 years of age. Early-onset Sjögren's syndrome poses diagnostic challenges, as classical sicca symptoms may be absent and extraglandular manifestations often predominate. This atypical presentation can result in delayed recognition, particularly in young women without overt glandular involvement (11).

There are reports of renal involvement in 5–14% of people with primary Sjögren's syndrome; tubulointerstitial nephritis is the most common symptom (12). Only 3–5% of patients have distal renal tubular acidosis, which is an uncommon renal symptom that rarely presents with hypokalemic paralysis (12,13). In many cases, renal manifestations

precede sicca symptoms by several years, as observed in this patient.

Women with anti-Ro/SSA and anti-La/SSB antibodies face a significant risk of immune-mediated fetal complications during pregnancy. Usually, between 12 and 16 weeks of gestation, transplacental antibody transfer causes the fetal atrioventricular node to become inflammatory and fibrotic, which leads to congenital heart block (14). The risk of CHB is estimated at 2–5% in a first anti-SSA–positive pregnancy and increases to 15–18% following a previously affected pregnancy (14). Once complete heart block develops, reversal is unlikely.

Hydroxychloroquine is now considered the cornerstone of management in anti-SSA–positive pregnancies. When started preconceptionally or early in pregnancy, HCQ has been demonstrated to decrease interferon-mediated inflammation and reduce the recurrence risk of congenital heart block by about 50% (13,15). Current rheumatology and obstetric guidelines recommend continuation of HCQ throughout pregnancy due to its favorable maternal and fetal safety profile (15).

Recent literature emphasizes that pregnancies complicated by Sjögren's syndrome are also associated with placental dysfunction, fetal growth restriction, hypertensive disorders of pregnancy, preterm birth, thromboembolic events, and increased cesarean delivery rates, even in the absence of fetal cardiac involvement (16). Accordingly, management requires a multidisciplinary approach with intensive fetal cardiac surveillance between 16 and 24 weeks of gestation, along with continued obstetric monitoring for placental insufficiency in the third trimester (16). The contrasting outcomes in this patient's two pregnancies underscore the critical importance of hydroxychloroquine adherence and structured fetal surveillance.

CONCLUSION

This case demonstrates that although anti-SSA–positive pregnancies carry a significant risk of fetal cardiac morbidity, outcomes can be substantially improved through preconception optimization, early and consistent hydroxychloroquine use, intensive fetal cardiac monitoring, and vigilant obstetric surveillance for FGR and Doppler abnormalities. Proactive obstetric and fetal medicine management resulted in a healthy neonatal outcome following a previously affected pregnancy.

Clinical Significance

This case emphasizes that primary Sjögren's syndrome can present at a young age with isolated extraglandular manifestations such as distal renal tubular acidosis, even in the absence of classical sicca symptoms, highlighting the need for early diagnostic vigilance. The contrasting outcomes of two consecutive pregnancies in the same patient demonstrate that adverse fetal cardiac outcomes may be preventable through preconception counseling, strict adherence to hydroxychloroquine therapy, and structured fetal cardiac surveillance. This report explains the importance of a multidisciplinary approach, integrating obstetric, fetal medicine, rheumatology, and nephrology care, to optimize maternal health and improve perinatal outcomes in high-risk autoimmune pregnancies.

LIST OF ABBREVIATIONS

- pSS – Primary Sjögren's syndrome
- SSA – Sjögren's syndrome–related antigen A
- SSB – Sjögren's syndrome–related antigen B
- Anti-Ro – Anti-Ro/SSA antibodies
- Anti-La – Anti-La/SSB antibodies
- RTA – Renal tubular acidosis
- dRTA – Distal renal tubular acidosis
- CHB – Complete heart block
- HCQ – Hydroxychloroquine
- FGR – Fetal growth restriction
- LSCS – Lower-segment cesarean section
- AV – Atrioventricular
- FHR – Fetal heart rate
- ABG – Arterial blood gas
- ANA – Antinuclear antibody
- TSH – Thyroid-stimulating hormone

Informed Consent: A written consent was taken from the patient to share clinical details related to the management of the case.

Conflict of Interest: None

REFERENCES

1. Bowman SJ. Primary Sjögren's syndrome. Lupus. 2018;27(Suppl 1):32–35.
2. Gupta S, Gupta N. Sjögren syndrome and pregnancy: a literature review. Perm J. 2016;21:16-047.
3. Bedei IA, Kniess D, Keil C, Wolter A, Schenk J, Sachs UJ, et al. Monitoring of women with anti-Ro/SSA and anti-La/SSB antibodies in Germany—status quo and intensified monitoring concepts. J Clin Med. 2024;13(4):1142.
4. Pruetz JD, Miller JC, Loeb GE, Silka MJ, Bar-Cohen Y, Chmait RH. Prenatal diagnosis and management of congenital complete heart block. Birth Defects Res. 2019;111(8):380.
5. Izmirly P, Kim M, Friedman DM, Costedoat-Chalumeau N, Clancy R, Copel JA, et al. Hydroxychloroquine to prevent recurrent congenital heart block in fetuses of anti-SSA/Ro-positive mothers. J Am Coll Cardiol. 2020;76(3):292–302.

6. Tan Z, Shao M, Zhou Y, Wang L, Ma Y, Xiang N, et al. Increased risk of adverse gestational outcomes in pregnant women with primary Sjögren's syndrome. RMD Open. 2024;10(2):e003616.
7. Izmirly PM, Costedoat-Chalumeau N, Pisoni CN, Khamashta MA, Kim MY, Saxena A, et al. Maternal use of hydroxychloroquine is associated with a reduced risk of recurrent anti-SSA/Ro associated cardiac manifestations of neonatal lupus. Circulation. 2012;126(1):76.
8. Berman A, Kenet G, Lubetsky A, Agmon-Levin N, Soffer S, Barg AA, et al. Hydroxychloroquine and pregnancy outcomes in patients with anti-phospholipid syndrome: a systematic review and meta-analysis. RMD Open. 2025;11(3):e005825.
9. Tesoriero L, Kidd J, Piccione J, Izmirly P, Akerman M, Carsons S, et al. Adverse pregnancy outcomes in Sjogren's disease compared to controls: an interdisciplinary approach with maternal–fetal medicine. AJP Rep. 2025;15(2):e47.
10. Freud LR, Simpson LL. Fetal cardiac screening: 1st trimester and beyond. Prenat Diagn. 2024;44(6-7):679–687.
11. Reed G, Deeb M, Mathew J, Rigby K, Cravens E, Raker C, et al. Pregnancy outcomes from a multidisciplinary obstetric-medicine/rheumatology clinic in the United States: a five-year retrospective analysis. Arthritis Care Res (Hoboken). 2024;76(12):1744–1750.
12. Sandhya P. Management of Sjögren's syndrome during pregnancy. In: Women's Health in Autoimmune Diseases. 2020. p. 187–196.
13. Vanoni F, Lava SAG, Fossali EF, Cavalli R, Simonetti GD, Bianchetti MG, et al. Neonatal systemic lupus erythematosus syndrome: a comprehensive review. Clin Rev Allergy Immunol. 2017;53(3):469–476.
14. Barsalou J, Costedoat-Chalumeau N, Berhanu A, Fors-Nieves C, Shah U, Brown P, et al. Effect of in utero hydroxychloroquine exposure on the development of cutaneous neonatal lupus erythematosus. Ann Rheum Dis. 2018;77(12):1742–1749.
15. Brucato A, Cimaz R, Caporali R, Ramoni V, Buyon J. Pregnancy outcomes in patients with autoimmune diseases and anti-Ro/SSA antibodies. Clin Rev Allergy Immunol. 2011;40(1):27–41.
16. Russell MD, Dey M, Flint J, Davie P, Allen A, Crossley A, et al. British Society for Rheumatology guideline on prescribing drugs in pregnancy and breastfeeding: immunomodulatory anti-rheumatic drugs and corticosteroids. Rheumatology (Oxford). 2023;62(4):e48–e88.
17. Cue LV, Rosenn B. An update on the approach to treatment of Sjogren's disease in pregnancy. J Matern Fetal Neonatal Med. 2024;37(1).

