# Obs and Gynae PG Focus Series Autoimmune Disorders in Pregnancy

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### Sjögren's Syndrome in Pregnancy

CHAPTER

#### INTRODUCTION

Sjögren's syndrome is an autoimmune disease with a chronic course, mainly affecting glands of the mouth and eyes. It can either be primary Sjögren's syndrome (pSS), which will present alone or it can be present with concomitant connective tissue diseases like systemic lupus erythematosus or rheumatoid arthritis.<sup>1</sup>

Sjögren's syndrome presents a wide range of clinical symptoms, from mild dryness of the mucosal surfaces to severe systemic involvement. The hallmark symptom of mucosal dryness results from immune-mediated inflammation that impairs the function of secretory glands.<sup>2</sup> While sicca features, such as dry eyes and mouth, significantly impact quality of life, systemic involvement is a key determinant of the disease's prognosis.<sup>3</sup> Sjögren's syndrome predominantly affects women, who are more likely to experience complicated pregnancies compared to women without the disease.<sup>4,5</sup>

#### PATHOPHYSIOLOGY

Laboratory diagnosis of Sjögren includes the following antibodies:

- Antinuclear antibodies (ANA)  $\rightarrow$  Most often detected
- Anti-SS-A (anti-Ro)  $\rightarrow$  This is the most specific antibody
- Anti-SS-B (anti-La)
- Cryoglobulins and hypocomplementemia → They are important prognostic markers<sup>6</sup>

In women with Sjögren's syndrome, certain markers can cause tissue damage, leading to pregnancy complications. These antibodies can cross the placenta around 12 weeks of gestation and potentially harm fetal tissues in several ways, including:

- Causing inflammation of the heart muscle (myocarditis)
- Disrupting the normal clearance of dying cells, which can lead to an abnormal immune response
- Triggering abnormal heart rhythms (arrhythmia)<sup>7,8</sup>

#### EFFECT OF PREGNANCY ON SJÖGREN'S SYNDROME

Pregnancy may worsen Sjögren's syndrome. Worsening in the postpartum period can be attributed to the development of pulmonary hypertension, which is known to worsen in the antennal and postnatal period.

#### EFFECT OF SJÖGREN'S SYNDROME ON PREGNANCY

Women with Sjögren's syndrome are at a higher risk of experiencing pregnancy complications compared to those without the disease. Although research on pregnancy outcomes in women with Sjögren's syndrome is limited, existing studies suggest an increased rate of:

- Spontaneous abortion
- Fetal loss<sup>9-11</sup>

These complications may be attributed to two factors:

- 1. Advanced maternal age at conception
- 2. Immunologic factors that contribute to miscarriage mechanisms

#### MANAGEMENT

Multidisciplinary team management involves an obstetrician who specializes in high-risk pregnancies, a rheumatologist, and a pediatrician.

#### **Prenatal Management**

It is important to counsel patient about the high risk of pregnancy and inform about the risks and complications involved. Optimizing the patient before pregnancy, so as to have a good disease control, minimum 3–6 months before conception is crucial.

#### **Antenatal Management**

One of the most serious complications of pregnancy in patients with Sjögren's syndrome is congenital heart block (CHB). Women who are anti-SS-A positive are at risk of having a baby with CHB. To manage this risk, frequent monitoring with serial echocardiograms and obstetric sonograms is necessary, starting between 16 and 20 weeks of gestation and continuing throughout the pregnancy.

The goal is to diagnose early and early treatment of incomplete CHB, thus improving the outcome for the fetus.<sup>12</sup> The rationales for management are—(1) to decrease transplacental transfer of antibodies, decreasing the disease severity by decreasing the antibodies and (2) to prevent occurrence of irreversible CHB, avoiding inflammation is important.<sup>13</sup> Although there is no conclusive evidence that steroids can reduce the levels of anti-SS-A or anti-SS-B antibodies or reverse CHB, research suggests that dexamethasone may be beneficial in certain situations. Dexamethasone has been shown to reverse carditis and incomplete CHB, as well as improve fetal heart function. Therefore, treatment with dexamethasone is recommended in cases where the CHB is recent or incomplete, or if there are signs of cardiac failure.<sup>14,15</sup>

Alternative therapies like plasmapheresis, intravenous immunoglobulins, and  $\beta$ -sympathomimetics can also be tried.<sup>15,16</sup> The best treatment option for a CHB would be implantation of a pacemaker after birth.

#### CONCLUSION

Sjögren's syndrome is associated with more complications during the antenatal and postnatal period. Preconceptional counseling, thus, can aid optimization to curb complications. This syndrome is best managed by a multidisciplinary approach involving a high-risk obstetrician, a rheumatologist, and a neonatologist.

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### **Obs and Gynae PG Focus Series Autoimmune Disorders in Pregnancy**

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