Vol. 01 Num. 03 (2025) 274–279

ANESTHETIC CONSIDERATIONS FOR CESAREAN DELIVERY IN PARTURIENT WITH SYSTEMIC SCLERODERMA COMBINING PULMONARY FIBROSIS, SEVERE SKIN INVOLVEMENT AND CREST SYNDROME

Rahmoune Farid Chakib[a]*, Lahoud Mari-José[b], Benhamou Dan[c]

[a]Hôpital privé du pays d'Auge, 175 rue Roger Ainsi, 14100 Lisieux, France.
[b]Hôpitaux Universitaires de Genève , Rue Gabrielle-Perret-Gentil 4, 1205 Genève, Suisse.
[c] Département d'Anesthésie Réanimation, CHU Bicêtre, 78, Rue du Général Leclerc, 94275 , Le Kremlin Bicêtre, Paris, France

[*] *Corresponding authors: Rahmoune Farid Chakib*
E-mail address: rahmounefc@yahoo.fr

Summary

Scleroderma or systemic sclerosis (SSC) is an autoimmune disease characterized by excessive accumulation of connective tissue components. Multisystem involvement of organs and pregnancy induced modifications can impact every aspect of anesthetic management and are causes of significant maternal morbi-mortality.
We report and discuss the peripartum management for cesarean delivery of a pregnant with SSC combining pulmonary fibrosis, severe skin involvement and CREST Syndrome.

Keywords : systemic scleroderma, cesarean delivery, pulmonary fibrosis, CREST syndrome

Citation: To be added by editorial staff during production.

Academic Editor: First name Last name

Received: date:02/02/2025
Revised: date:04/03/2025
Accepted: date:14/06/2025
Published: date:22/06/2025

1. Introduction :

Scleroderma or systemic sclerosis (SSC) is an autoimmune disease characterized by fibrosis affecting the skin with multisystem involvement of organs (heart, lungs, gastrointestinal tract and kidneys).
Women are affected more commonly than men, especially during the reproductive years [1]. Pregnancy worsens the severity profile of scleroderma in approximately 20% of parturient [2]. Multisystem involvement and impact of pregnancy are causes of significant maternal morbi-mortality.
Some case reports have already presented anesthetic care of these patients but in a few of them were pregnant women with multisystem disease described. We report and discuss the anesthetic management for cesarean delivery in a parturient with SSC combining pulmonary fibrosis, severe skin involvement and CREST Syndrome.

2. Case description

A 31-year-old woman with systemic scleroderma (SSC) was oriented to our maternity unit at 24 weeks of gestation for her first pregnancy because of her disease and associating with fetal anomalies (breech presentation and diaphragmatic hernia). She had been diagnosed with SSC seven years ago, and clinical features present at diagnosis were essentially skin with widespread cutaneous sclerosis. During her clinical history, she had vascular lesions manifested by CREST syndrome and chronic respiratory failure due to a pulmonary fibrosis. Her Raynaud's phenomenon was primarily treated with a calcium channel blocker and sildenafil stopped at announcement of pregnancy

Proximal muscle weakness due to myositis was also present. Investigations for immune antibodies were negative except for antinuclear antibodies. Because of difficulties implementation of peripheral venous access, she had an implantable central venous catheter one year ago. During her disease, she had been treated with immunosuppressant drugs (methotrexate, immunoglobulins and steroids) and all these treatments were stopped during pregnancy after a coordinated multidisciplinary meeting including obstetricians, rheumatologists and neonatologists.

Clinical examination on admission showed mild dyspnea (NYHA stage 3) with oxygen saturation at 94% on air breathing, no signs of hemodynamic or cardiac failure. Anesthetic evaluation showed a high risk of difficult airway management with a grade IV Mallampati class, very restricted mouth opening (2 cm) and limited cervical extension. No telangiectasias were seen on lips, tongue or mouth mucosa. Spinous processes were easily palpable. Difficulties in obtaining venous access were present, but the catheter implanted a year ago was available.

Investigations to define disease severity showed an interstitial lung disease associated to a mild restrictive syndrome with FEV1 52%/ 2.17L, and pulmonary function tests showed decreased compliance and an altered diffusion capacity (DLCO = 34%).Transthoracic echocardiography was normal (left ventricular ejection fraction 69%, no pericardial infusion or pulmonary arterial hypertension, and normal right values). The patient had no renal or liver laboratory abnormalities. Hematologic tests were normal except for a decreased factor XII concentration with no bleeding risk.

A multidisciplinary staff meeting concluded that there was a significant risk of failure to progress in labor because of probable vaginal constriction and breech presentation, and decided to perform an elective cesarean delivery at 38 weeks under neuraxial anesthesia because of her high risk of aspiration and likely difficult airway management. However, at 36 weeks of gestation, she presented fever and dyspnea. Urinary and blood culture samplings remained negative. Nicardipine and labetolol were necessary to control blood pressure which increased up to 190/110 with urinary protein level at 1,7 g /L concluding to recent pre-eclampsia. Large spectrum antibiotic therapy was started (ceftriaxone, gentamycine and metronidazole). Decision to perform caesarean delivery was taken after normal body temperature and blood pressure (138/86) were obtained without worsening pre-eclampsia.

A combined spinal epidural anesthesia was performed at L2-L3 space with mild technical difficulties due to obstacles in positioning the patient. An intrathecal mixture of hyperbaric bupivacaine 8 mg with sufentanil 3 µg and morphine 0.1 mg was injected. Carbetocine 100 µg was slowly infused after extraction. Apgar scores were 8/10/10 at 1, 5 and 10 min and the newborn was transferred to the neonatal intensive care unit for taking care of his hernia. Additional 10 mL of lidocaine via the epidural catheter were necessary to maintain an adequate quality of anesthesia. In the operative room, close attention was given to the patient positioning and body temperature. No major hypotension was noted, and nasal oxygen was maintained for oxygen saturation upper than 95%. The anesthetic and surgical procedures were uneventful and parturient was transferred to the obstetrical high dependency unit. She remained stable under oral nicaridpine, no clinical or biological manifestation of pre-eclampsia or HELLP syndrome and he remained eupneic with no oxygen needed. On post-operative day 2, two blood cultures drawn from the catheter were positive with methicillin-sensitive staphylococcus aureus. antibiotic therapy was adapted. Chamber infection was considered leading to device withdrawal. The patient remained stable with no disease exacerbation and was discharged 8 days postoperatively.

3. Discussion

Systemic scleroderma (SSC) is a connective tissue disease with diffuse sclerosis and inflammation affecting skin, joints, blood vessels, heart, lungs, gastrointestinal tract and kidneys **[1]**. One particular combination is the CREST syndrome (acronym of Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasia). The disease has a multifactorial origin including vascular dysfunction, inflammation and connective tissue fibrosis **[3]**. It has a prevalence ranging from 5-490 cases per million with an incidence of 2–10 cases per million per year and a peak incidence in the fourth decade of life **[4]**. Women are more commonly affected than men (ratio of 4–10:1) which increases during the reproductive years **[5-6]**, but there is no decrease in overall fertility **[7]**. Spontaneous abortion, stillbirth and prematurity are more often encountered due to placental vasculopathy **[8]**. Overall, pregnancy worsens evolution of scleroderma in approximately 20% of parturient, with stabilization in 50% of cases and improvement of symptoms in others **[2]**. There is no reported aggravation of skin disease during pregnancy, but organ involvement may worsen **[9]**. Some case reports have already presented perioperative management of patients with SSC **[2, 10-11]**, but anesthetic management of pregnant women for vaginal or cesarean delivery with severe SSC associating CREST syndrome, severe skin involvement and pulmonary fibrosis has rarely been described.

In over 70% of patients with SSC, pulmonary manifestations develop and range between mild interstitial lung disease secondary to pulmonary infiltration and pulmonary hypertension with right-sided heart failure due to vasculopathy **[12]**. Pulmonary fibrosis (PF) with severe restrictive syndrome (FEV1< 30%), chronic respiratory failure or severe pulmonary hypertension has a prevalence is estimated between 20 and 100% during the course of the disease and its occurrence forbids pregnancy **[12-13]**. The PF is secondary to fibroblast proliferation and collagen secretion induced by IL-4 secretion and INF-1 inhibition **[14]**. Cytokine deregulation is produced by epithelial and endothelial destruction by inflammation **[14]**.

Our parturient had worsened her pulmonary disease with an increase of her FEV1 for 70% (before pregnancy) till 52%, with dyspnea stage 3 of NYHA. . Indeed, with pregnancy-induced increase in blood volume, heart rate and cardiac output, women frequently complain of worsening shortness of breath with increasing gestation **[15]**. The decreases in functional residual capacity and closing volume can lead to rapid desaturation and hypoxemia / hypercapnia which may exaggerate pulmonary vascular resistance **[16-17]**. Mechanical ventilation may be difficult secondary to reduced compliance and positive pressure ventilation may decompensate hemodynamic stability in patients with pulmonary hypertension **[18-19]**.

Cardiac complications of scleroderma are mostly secondary to pulmonary hypertension but primary cardiac lesions may exist **[20]**. "Myocardial Raynaud's phenomenon" manifests as a subclinical chronic ischemia due to myocardial microcirculation abnormalities. Ventricular hypertrophy, diastolic dysfunction, pericarditis and arrhythmias may occur **[20]**. Hemodynamic changes in pregnancy worsen cardiac abnormalities with increased risk of arrhythmias and potential heart failure **[21]**. Our parturient did not present with cardiac manifestations of SSC and echocardiographic explorations were normal.

Maintenance of preload is important especially if neuraxial anesthesia is considered given the risk of hemodynamic worsening secondary to vasoplegia. Our parturient had a mild restrictive syndrome with FVC around 50%. Pregnancy was risky because rapid deterioration might have occurred especially in the third trimester. Owing to the usual benefit in pregnant women and to the risk of respiratory complications, regional anesthesia with maintenance of spontaneous ventilation

was preferred. Recent case reports of neuraxial anesthesia using bupivacaine, ropivacaine and prilocaine have not reported any block prolongation **[17]**.

Renal crisis is a serious complication of SSC and the primary cause of maternal death in scleroderma pregnancies. It is characterized by acute renal failure, malignant hypertension and proteinuria **[14]**. Pathophysiology of renal lesions include thickening of renal glomerular basement membrane, hyperplasia of interlobular arterioles **[17]**. Microangiopathic hemolytic anemia with thrombocytopenia may coexist **[15]**. The major risks identified are patients with recent onset disease and diffuse, rapidly progressive skin involvement. Pregnancy itself has been hypothesized to precipitate renal crisis, though no increased incidence of renal crisis in pregnancy has been demonstrated **[7, 22]**. Hypertension and proteinuria in preeclampsia can be easily confounded with renal crisis. However, no liver function perturbation is associated. ACE inhibitors, despite their known foetotoxicity, should be prescribed if renal crisis occurs because it is the only treatment shown to reduce mortality **[23]**. Anesthetic management includes modified drug dosage because of decreased renal clearance. In our case, the patient presented with preeclampsia associating hypertension and proteinuria with normal renal and liver functions. Hypertension was rapidly controlled before decision of cesarean and was persistent 3 weeks after delivery without any other clinical or biological anomalies.

In SSC, skin fibrosis may affect hands, face, arms and upper chest. Pregnancy is not known to worsen skin disease. Fibrosis involving face and neck may cause restricted mouth opening and limit neck extension making (like in our patient) mask ventilation and tracheal intubation potentially difficult **[17-18]**. Airway management in the parturient may be even more difficult if associated with edema, preeclampsia or obesity. The cutaneous shrinkage seen in diffused scleroderma may affect abdominal wall and may induce a risk of abdominal compression with a gravid uterus. Skin fibrosis may increase technical difficulty when performing neuraxial anesthesia because patient positioning may cause restricted joint mobility. Positioning in operation room was meticulous with use of extra padding . Hand and arm lesions may lead to flexion contracture, hypoperfusion ulceration with hands deformities **[24]**. Difficulty in obtaining venous access is frequent due to dermal thickening and central venous access may be necessary (like in this case). Non-invasive blood pressure and pulse oximeter monitoring may be compromised by skin fibrosis and vasoconstriction **[24]**. Hypothermia may indeed trigger vascular crisis and overheating with sweating process hindered in patients with scleroderma, may manifest as malignant hypertension **[25]**. In our patient, pulse oximeter was changed from a finger to another to avoid ischemic damage. Special warming of the operation room and a thermal blanket was used to avoid body temperature fluctuation.

In patients with SSC, vascular manifestations can be exhibited by the CREST syndrome associating finger Calcinosis, Raynaud’s syndrome, Esophageal dysmotility with chronic gastro-esophageal reflux and major esophageal distension (GERD), Sclerodactyly, and Telangiectasia. Raynaud's phenomenon tends to improve in pregnancy secondary to generalized peripheral vasodilatation and increased cardiac output. Tongue and mouth mucosa telangiectasias are prone to bleeding and careful airway manipulation is required **[2, 18]**. Hyperemia in pregnancy may trigger telangiectasia bleeding elsewhere and gastrointestinal bleeding may occur secondary to gastric lesions known as the "estomac pastèque" **[14]**. Esophageal dysmotility, GERD and delayed gastric emptying caused by mucosal shrinkage and exacerbated by the gravid uterus in the parturient with SSC may increase risk of aspiration during general anesthesia **[24]**. Coagulopathy secondary to vitamin K malabsorption may be an obstacle to performing a neuraxial procedure. In patients with scleroderma, vascular spasm may involve placental arteries resulting in miscarriage and intrauterine growth restriction but the

frequency of premature infants was identical to parturient with rheumatoid arthritis, with a slight increase compared to general population **[26]** résultats.

Conclusion

In summary, SSC can pose a significant challenge for anesthetists especially in pregnant women. Multisystem involvement of organ and impact of pregnancy must be evaluated to ovoid complications. Anesthesiologists managing parturient in peri-partum with SSC must have experience and background about the pathogenesis, clinical manifestations, systemic involvement and anesthetic considerations. Regional anesthesia must be preferred because of predicted difficulties of airway management and high risk of general anesthesia especially if pulmonary or cardiac involvements and CREST Syndrome are present. In the operative room, special care must be applied to ovoid ischemic damage and temperature fluctuations. In pregnancy, careful pre-partum assessment is required to determine the multisystem involvement of systemic sclerosis and allow formulation of an anesthetic decisions.

Declaration of interests : The authors have nothing to declare.

References

[1] Eisele JH: Connective tissue diseases. In Anesthesia & Uncommon Diseases 4th edition. Edited by: Benumof JL. Philadelphia, USA: Saunders Company; 1998:399-421.

[2] Thompson J, Conklin KA. Anesthetic management of a pregnant patient with scleroderma. Anesthesiology1983 ;59(1):69-71.

[3] Roberts JG, Sabar R, Gianoli JA, Kaye AD. Progressive systemic sclerosis: clinical manifestations and anesthetic considerations. J Clin Anesth 2002;14(6):474-7.

[4] Chifflot H, Fautrel B, Sordet C, Chatelus E, Sibilia J. Incidence and prevalence of systemic sclerosis: a systematic literature review. Semin Arthritis Rheum 2008 ;37(4):223-35.

[5] Barnes J, Mayes MD. Epidemiology of systemic sclerosis: incidence, prevalence, survival, risk factors, malignancy, and environmental triggers. Curr Opin Rheumatol. 2012 ; 24(2):165-70.

[6] Steen VD. Pregnancy in scleroderma. Rheum Dis Clin North Am 2007;33(2):345-58.

[7] Ballou SP, Morley JJ, Kushner I. Pregnancy and systemic sclerosis. Arthritis Rheum 1984; 27(3):295–8.

[8] Doss BJ,Jacques SM, Mayes MD, Qureshi F. Maternal scleroderma: placental findings and perinatal outcome. Hum Pathol 1998 ;29(12):1524-30.

[9] Miniati I, Guiducci S, Mecacci F, Mello G, Matucci-Cerinic M. Pregnancy in systemic sclerosis.Rheumatology (Oxford). 2008 ;47, 3:16-8.

[10] Bailey AR, Wolmarans M, Rhodes S. Spinal anaesthesia for caesarean section in a patient with systemic sclerosis. Anaesthesia 1999;54(4):355-8.

[11] Ye F, Kong G, Huang J. Anesthetic management of a patient with localised scleroderma. Springerplus. 2016 Sep 7;5(1):1507.

[12] Bonnin M, Mercier FJ, Sitbon O, Roger-Christoph S, Jaïs X, Humbert M, Audibert F, Frydman R, Simonneau G, Benhamou D. Severe pulmonary hypertension during pregnancy: mode of delivery and anesthetic management of 15 consecutive cases. Anesthesiology 2005;102(6):1133-7.

[13] Schurawitzki H, Stiglbauer R, Graninger W, Herold C, Pölzleitner D, Burghuber OC, Tscholakoff D. Interstitial lung disease in progressive systemic sclerosis: high-resolution CT versus radiography. Radiology 1990;176(3):755-9.

[14] Bégneu E, Aïssou M, Lescot T, Cabane JP, Beaussier M. Perioperative management of patients with systemic scleroderma. Ann Fr Anesth Reanim 2014;33(12):669-76.

[15] Lidar M, Langevitz P. Pregnancy issues in scleroderma. Autoimmun Rev. 2012 ;11(6-7):A515-9.

[16] Monnery L, Nanson J, Charlton G. Primary pulmonary hypertension in pregnancy; a role for novel vasodilators. Br J Anaesth 2001;87(2):295-8.

[17] Dempsey ZS, Rowell S, McRobert R. The role of regional and neuroaxial anesthesia in patients with systemic sclerosis. Local Reg Anesth 2011;4:47-56.

[18] Kanter GJ, Barash PG. Undiagnosed scleroderma in a patient with a difficult airway. Yale J Biol Med 1998 J;71(1):31-3.

[19] Goland S, Tsai F, Habib M, Janmohamed M, Goodwin TM, Elkayam U. Favorable outcome of pregnancy with an elective use of epoprostenol and sildenafil in women with severe pulmonary hypertension. Cardiology 2010;115(3):205-8.
[20] Champion HC. The Heart in Scleroderma. Rheum Dis Clin North Am. 2008 February ; 34(1): 181–viii.
[21] Chiang CH, Liu CJ, Huang CC, Chan WL, Huang PH, Chen TJ, Chung CM, Lin SJ, Chen JW, Leu HB. Systemic sclerosis and risk of ischaemic stroke: a nationwide cohort study. Rheumatology (Oxford) 2013; 52(1):161-5.
[22] Steen VD. Pregnancy in women with systemic sclerosis. Obstet Gynecol 1999;94(1):15-20.
[23] Steen VD, Costantino JP, Shapiro AP, et al. Outcome of renal crisis in systemic sclerosis: relation to availability of angiotensin converting enzyme (ACE) inhibitors. Ann Intern Med 1990;113(5):352–7.
[24] Roberts JG, Sabar R, Gianoli JA, Kaye AD. Progressive systemic sclerosis: clinical manifestations and anesthetic considerations. J Clin Anesth 2002;14(6):474-7.
[25] Smoak LR. Anesthesia considerations for the patient with progressive systemic sclerosis (scleroderma). AANA J 1982;50(6):548–554.
[26] Josselin-Mahr L, Carbonne B, Cabane J. Systemic sclerosis and pregnancy. Rev Med Interne. 2011 Jun;32(6):363-8.