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Selected Reports

Restrictive Lung Disease in Pregnancy

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Restrictive lung disease is uncommon in pregnancy. We reviewed 15 pregnancies in 12 women with restrictive disease due to kyphoscoliosis, neuromuscular disease, or parenchymal lung disease. Median FVC was 40% predicted, and six women (50%) had an FVC < 1.0 L. In the 14 pregnancies in which at least two spirometry readings were available, FVC increased in three pregnancies, decreased in three, and remained stable in eight, with maximal changes of 0.4 L. Three women required supplemental oxygen, and one woman with neuromuscular disease required noninvasive ventilation. Premature delivery occurred in nine pregnancies (60%), and 10 deliveries (67%) were by cesarean section. Neuraxial anesthesia was used in 10 of 15 deliveries but was limited in the others by difficult spinal anatomy. There was no maternal or neonatal mortality. Women with restrictive lung disease tolerate pregnancy reasonably well, but many have premature delivery. A multidisciplinary approach is essential, with monitoring of spirometry and oxygenation and planning for labor and delivery.

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Abbreviations: NIV = noninvasive ventilation; SLE = systemic lupus erythematosus

Restrictive lung disease may result from a parenchymal interstitial process or from a chest wall or neuromuscular abnormality. Interstitial lung disease is relatively uncommon in pregnancy, because most of these conditions have a time of onset after the reproductive years.[1] Few data exist to inform the risks of pregnancy in women with restrictive lung disease or to plan for appropriate peripartum care. Pregnancy is associated with physiologic changes, including a reduction in lung volumes[2] and an increased ventilatory requirement, which may be problematic in women with preexisting restrictive disease.[3] Furthermore, interstitial conditions may affect gas transfer and oxygenation, putting the fetus at risk of hypoxia. A previous case series of pregnant women with interstitial and chest wall/neuromuscular lung disease described nine cases with various levels of severity, but with good outcomes.[4] However, some literature suggests that women with severe restrictive lung disease (FVC < 1.0 L) should avoid pregnancy because of the maternal risks.[1]

The goal of this study was to review our experience with the management of pregnant patients with severe restrictive lung disease. Specifically, we describe the characteristics of these patients, and maternal and fetal outcomes.

Materials and Methods

We retrospectively reviewed the charts of women with restrictive lung disease who were followed in our high-risk pregnancy program between 2001 and 2012. Mount Sinai Hospital is a tertiary-care referral center with 6,500 deliveries annually and has a level 3 neonatal and adult ICU. Patients were identified from an ICU database and from respirologists' and maternal-fetal medicine specialists' office charts. The study was approved by the Mount Sinai Hospital Research Ethics Board, and the need for informed consent was waived (Approval No. 11-0316-C).

Pregnant women with a documented FVC of < 70% predicted were evaluated for the study; patients with predominant obstructive airways disease were excluded. Data extracted from charts included demographic data, preexisting conditions, pulmonary function tests (prepregnancy and all available during pregnancy), pregnancy course and requirement for respiratory support, labor and delivery, and maternal and fetal outcomes. Where available, the earliest spirometry during pregnancy was compared with the last during that pregnancy; an increase or decrease in FVC of > 10% was considered significant.

Results

We identified 12 patients who underwent 15 pregnancies. Mean maternal age was 30.3 years (SD, 5.1 years). Restrictive disease was due to chest wall conditions (n = 6), such as

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Some of these data have been presented previously at the 6th Meeting of the International Society for Obstetric Medicine, July 7-8, 2012, London, England, and at CHEST 2012, October 20-25, 2012, Atlanta, GA, and have been presented in abstract form (Tram C, Lapinsky S, Maxwell C. Restrictive lung disease in pregnancy. *Chest*. 2012;142[4_MeetingAbstracts]:382A).

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kyphoscoliosis and osteogenesis imperfecta; neuromuscular diseases (n = 2) (namely, congenital myasthenia gravis and spinal muscular atrophy, both with associated kyphoscoliosis); and parenchymal lung disease (n = 4) due to scleroderma, systemic lupus erythematosus (SLE), bronchiectasis, and chronic granulomatous disease with recurrent invasive *Aspergillus* infection. A transthoracic echocardiogram was obtained in 12 pregnancies; the women with scleroderma and SLE demonstrated mild pulmonary hypertension (right ventricular systolic pressure of 52 mm Hg and 40 mm Hg, respectively). No other significant comorbidities were noted in this patient group. No patients were receiving specific therapy for interstitial lung disease during the pregnancy period.

Spirometry

At least two spirometry readings were available in 14 of the 15 pregnancies. The earliest spirometry was from 7 to 31 weeks' gestation (median, 17 weeks) and the last in the pregnancy at 23 to 37 weeks (median, 31 weeks). Initial FVC values are given in Table 1; six women (50%) recorded an initial FVC of < 1.0 L. Two patients, one with bronchiectasis and one with scoliosis, had evidence of associated obstructive airways disease, with FEV_1/FVC ratios of 67% and 63%, respectively. The remaining 10 patients had a median FEV_1/FVC ratio of 89%. During pregnancy, FVC decreased in three pregnancies (maximal decrease, 300 mL), remained stable in eight pregnancies, and increased in three pregnancies (maximal increase, 400 mL). The change in serial spirometry did not appear to be associated with the underlying disease process (Fig 1).

Because of the retrospective nature of this study, lung function tests were available only as performed by the patient's obstetrician or respirologist. As a result, not all patients had full lung function tests available (ie, several were missing total lung capacity and diffusing capacity), and few had serial spirometry or regular oxygenation assessments throughout the pregnancy. A minority of patients had prepregnancy spirometry, and the results obtained were very similar to those of the earliest spirometry in the pregnancy. Tests of respiratory muscle strength and exercise capacity were not performed.

Oxygenation

Oxygen saturation by pulse oximetry on room air during the pregnancies was in the range of 86% to 100% and was below 90% in only two patients; in one of these patients this occurred only during exercise. Three women were treated with supplemental oxygen during their four pregnancies (Table 2); one patient, with SLE, demonstrated transient desaturation during exercise during her first pregnancy and was started on oxygen empirically in a second pregnancy. The patient with congenital myasthenia gravis and scoliosis required intermittent oxygen therapy and then intermittent and nocturnal noninvasive ventilation (NIV) in the third trimester of both of her pregnancies. Her arterial blood gas results showed borderline hypercapnia, with readings of 36 to 40 mm Hg at a gestational age at which the expected $Paco_2$ range is 30 to 32 mm Hg.[5]

Delivery and Outcome

Only three pregnancies underwent spontaneous labor with vaginal delivery; the remaining 12 were induced or underwent elective cesarean section (Table 1). Nine pregnancies were delivered preterm, at gestations of 31 to 36 weeks (Table 1). The earliest preterm delivery at 31 weeks' gestation was due to preeclampsia in the patient with scleroderma.

Table 1—*Underlying Condition, Initial FVC During Pregnancy, and Details of Delivery*

Underlying Condition	FVC (% Predicted), L	Gestation at Delivery, wk	Mode of Delivery	Anesthesia
Parenchymal				
CGD	2.66 (68)	40	Vaginal[a]	Neuraxial
Scleroderma	1.9 (54)	31	C-S[b]	Neuraxial
SLE	1.9 (65)	35	Vaginal[a]	Neuraxial
	1.8 (62)	35	Vaginal[a]	Neuraxial
Bronchiectasis	0.9 (31)	39	Vaginal	Neuraxial
Chest wall				
Kyphoscoliosis	0.95 (29)	35	C-S	Neuraxial
Scoliosis	1.36 (44)	38	Vaginal	Neuraxial
Kyphoscoliosis and K-F	0.68 (20)	35	C-S	GA
Kyphoscoliosis	2.0 (58)	38	C-S[b]	GA
Kyphoscoliosis	1.0 (40)	38	C-S	Neuraxial
	0.9 (31)	38	C-S	GA
Osteogenesis imperfecta	1.16 (59)	35	C-S	GA
Muscular				
Congenital myasthenia	0.9 (30)	33	C-S	Neuraxial
	0.8 (22)	32	C-S	Neuraxial
Spinal muscular atrophy	0.95 (33)	36	C-S	GA

CGD = chronic granulomatous disease; C-S = cesarean section; GA = general anesthesia; K-F = Klippel-Feil syndrome; SLE = systemic lupus erythematosus.

[a] Spontaneous delivery.

[b] Failed induction.