

# Idiopathic Subglottic Stenosis during Pregnancy: A Support Group Survey

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## Abstract

**Objectives:** (1) To determine how pregnancy affects idiopathic subglottic stenosis (iSGS) symptoms. (2) To determine treatments utilized (including operating room (OR) and in-office procedures) for iSGS before, during, and after pregnancy.

**Methods:** A 24-question survey was distributed to an international iSGS support group to assess the patient experience among individuals who have been pregnant with diagnosis of iSGS. Descriptive statistics and chi<sup>2</sup> analyses were completed.

**Results:** A total of 413 iSGS patients participated; 84.7% (n = 350) of patients were diagnosed *prior* to menopause. A total of 25.5% patients reported being pregnant when they had airway stenosis; 71.1% of those reported more severe airway symptoms during pregnancy. The proportion of patients that reported requiring  $\geq 1$  OR interventions (microlaryngoscopy and bronchoscopy, laser, balloon dilation, or steroid injection) before, during and after pregnancy was the following, respectively: 37.3%, 35.6%, 51.3%. Whereas the proportion of patients that reported requiring  $\geq 1$  in-office interventions (awake balloon dilation or steroid injection) before, during and after pregnancy was the following, respectively: 13.6%, 11.8%, 15.8%. Number of pregnancies and age of diagnosis was not related to severity of symptoms or requiring more airway interventions ( $P > .05$ ).

**Conclusions:** This survey reveals worsening of symptoms during pregnancy, however, this did not lead to increase in operative or clinic interventions to improve airway symptoms during pregnancy. Future avenues for research include optimizing management of airway symptoms during pregnancy to limit OR-based interventions.

**Level of evidence:** Level IV

## Keywords

idiopathic subglottic stenosis, stridor, airway surgery, subglottic stenosis, survey

## Introduction

Subglottic stenosis is a rare life threatening fibroinflammatory disease that narrows the upper airway leading to severe dyspnea. The etiology of subglottic stenosis is varied and may include iatrogenic etiologies (prior intubation or tracheostomy), gastroesophageal reflux disease, inflammatory disorders including sarcoidosis and granulomatosis with polyangiitis (GPA), and idiopathic.<sup>1</sup> Of those diagnosed with subglottic stenosis, an estimated up to 20% are thought to have primary idiopathic subglottic stenosis (iSGS).<sup>2</sup> Among patients with iSGS, the vast majority are adult Caucasian women; most of these women are first diagnosed during their peri-menopausal years, but onset can occur anywhere between 20 and 70 years.<sup>3,4</sup>

Although the age of onset of iSGS can occur in child-bearing years, little research has been conducted to assess the effect of pregnancy on the disease process and how pregnancy affects the necessity for further treatments.

Certainly, this is concerning as worsening of iSGS during pregnancy can impair quality of life or even threaten the life of both the mother and fetus. Historically, iSGS has required multiple, periodic operative treatments including endoscopic dilation (rigid or balloon) and endoscopic resection (cautery or laser) with adjuvant medication delivered topically or intralesional, with symptomatic recurrence occurring on average 12.6 months after a procedure. However, this condition can ultimately be treated with open surgical

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resection and/or tracheostomy.<sup>5-7</sup> These types of treatments could pose unique difficulties during pregnancy, namely fetal safety with repeated anesthetic use.

Given the paucity of research regarding the treatment of iSGS during pregnancy, a survey study of an international iSGS support group was completed. The aim of this study was to better understand the patient experience among those diagnosed with iSGS during pregnancy. Furthermore, we aim to appreciate peripartum effects on the disease and symptoms progression, in addition to necessity of further treatments. iSGS pregnancy-related complications and attitudes toward future childbearing are also evaluated.

## Methods

This was a survey study conducted among an international, online iSGS support group (Living with Idiopathic Subglottic Stenosis; [www.facebook.com/groups/idiopathicsubglotticstenosis](http://www.facebook.com/groups/idiopathicsubglotticstenosis)). The iSGS support group is independently run through a social media platform (Facebook, [www.facebook.com](http://www.facebook.com); Menlo Park, CA) and currently contains 4030 members. Members include iSGS patients, in addition to patients' family members, physicians, nurses, students, and other medical support staff. A 24-question survey was distributed to members of the support group in November of 2019; the survey was left open for a total of 2 weeks. The survey was drafted by 3 physicians familiar with the diagnosis and treatment of iSGS (HM, VT, MS) and was emailed to the independent moderator (CA) who provided feedback and edits. The finalized survey consisted of the following types of questions: demographics (n=4), type and etiology of stenosis (n=3), queries regarding relationship between diagnosis of iSGS and pregnancy (n=3), types of treatments obtained during, before, and after pregnancy (n=7), assessment of airway symptoms in relation to pregnancy (n=2), complications during pregnancy (n=1), lifestyle effects (n=2), and effect of hormones on disease (n=2). The moderator then posted the survey to the iSGS support group using an online survey platform (SurveyMonkey, [www.surveymonkey.com](http://www.surveymonkey.com); San Mateo, CA). All current members with airway stenosis were eligible to provide responses; participation was voluntary and anonymous. Duplicate responses were prevented through adjusting the settings within SurveyMonkey. During the time the survey was open, a series of 3 reminders regarding survey completion were sent via email and Facebook. If patients had an etiology for their subglottic stenosis other than idiopathic (including autoimmune, polytrauma, or iatrogenic) they were excluded from analysis (n=20).

STATA/IC 14.0 was used to conduct data analysis. Demographic and treatment information was assessed using standard descriptive and frequency analyses. Chi<sup>2</sup> test was used for categorical variables to assess the relationship between number of pregnancies and age of diagnosis in relationship to disease severity and number of treatments

required. This protocol was granted exempt status by the University of Utah Institutional Review Board (IRB).

## Results

A total of 413 iSGS patients participated in the survey; all of these patients indicated their stenosis as being subglottic in nature and all patients were female. A majority of patients indicated they were diagnosed with iSGS *prior* to menopause (84.7%, n=350), with the most common age group for diagnosis being 35 to 44 years of age (32.2%, n=133). Some participants had been treated for their iSGS for >20 years (1.9%), but the majority of participants (51.6%) had undergone treatment for their disease between 1 and 5 years in length. See Table 1 for patient demographics.

When assessing frequency of pregnancies among this population, 20.9% (n=86) indicated they had never been pregnant. Among those who indicated they had been pregnant, 25.5% (n=83) patients reported being pregnant and having a diagnosis of iSGS; 28.9% (n=24) of those patients endorsed that their airway symptoms *first* appeared during pregnancy. In terms of airway symptoms during pregnancy, 71.1% (n=54) described their symptoms as *much worse* or *worse* while they were pregnant. Approximately 36.8% (n=28) had described their airway symptoms after pregnancy as *better* or *much better*, however, 36.8% (n=28) indicated *no change* in their airway symptoms after delivery. See Figures 1 and 2. Number of pregnancies and age of diagnosis was not related to severity of symptoms during/after pregnancy ( $P > .05$ ) or requiring more airway interventions during/after pregnancy ( $P > .05$ ).

The breakdown of patients requiring  $\geq 1$  operative interventions (microlaryngoscopy and bronchoscopy, laser, balloon dilation, or steroid injection) in the 12 months before pregnancy, during pregnancy, and in the 12 months after pregnancy was the following, respectively: 37.3%, 35.6%, 51.3%. Whereas the proportion of patients that reported requiring  $\geq 1$  in-office interventions (awake balloon dilation or steroid injection) in the 12 months before, during pregnancy, and in the 12 months after pregnancy was the following, respectively: 13.6%, 11.8%, 15.8%. See Figures 3 and 4. The most common pregnancy complication was preeclampsia, affecting 21.7% (n=18) of patients, followed by anemia 9.6% (n=8) and bedrest 8.4% (n=7). Approximately 40.8% (n=31) of patients stated their experience with iSGS adversely affected their decision to have children in the future.

When queried regarding the impact of oral contraceptives and post-menopausal hormones, there did not appear to be symptomatic worsening related to their use. Among those who endorsed using oral contraceptives (68.8% of participants), only 15% stated they *agree* or *strongly agree* that their symptoms became worse after starting oral contraceptives. While only 8.8% of participants had used

**Table 1.** Characteristics of Survey Participants.

| Characteristic                            | Result       |
|---|--------------|
| <b>Age of diagnosis</b>                   | <b>% (n)</b> |
| Pre-menopausal                            | 84.7 (n=350) |
| Post-menopausal                           | 15.3 (n=63)  |
| <b>Current age (years)</b>                | <b>% (n)</b> |
| 18-24                                     | 0.24 (n=1)   |
| 25-34                                     | 9.9 (n=41)   |
| 35-44                                     | 23.5 (n=97)  |
| 45-54                                     | 34.4 (n=142) |
| 55-64                                     | 22.8 (n=94)  |
| 65+                                       | 9.2 (n=38)   |
| <b># of years undergoing treatment</b>    | <b>% (n)</b> |
| <1  | 6.0 (n=25)   |
| 1-5                                       | 51.6 (n=213) |
| 6-10                                      | 24.5 (n=101) |
| 11-15                                     | 12.7 (n=52)  |
| 16-20                                     | 3.2 (n=13)   |
| >20                                       | 1.85 (n=7)   |
| <b># of times pregnant</b>                | <b>% (n)</b> |
| 0   | 20.9 (n=86)  |
| 1   | 12.1 (n=50)  |
| 2   | 30.3 (n=125) |
| 3   | 19.4 (n=80)  |
| 4   | 10.7 (n=44)  |
| ≥5  | 6.6 (n=27)   |
| <b>Pregnancy with history of stenosis</b> | <b>% (n)</b> |
| Yes                                       | 25.5 (n=83)  |
| No  | 74.5 (n=242) |

hormones after menopause, there were no participants who endorsed worsening of their symptoms with post-menopausal hormone use.

## Discussion

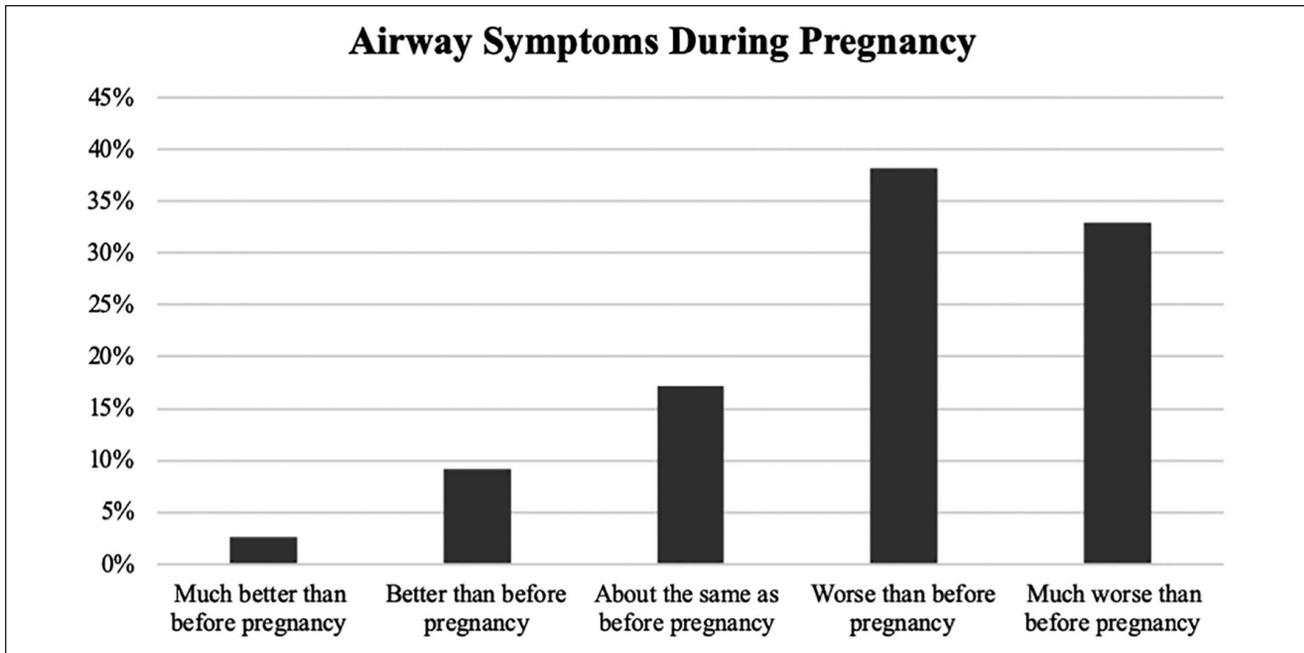
iSGS is an uncommon cause of upper airway obstruction, which nearly universally occurs in adult Caucasian women. The most common symptoms of iSGS include dyspnea on exertion, cough, wheezing, stridor, and difficulty clearing secretions.<sup>2,8</sup> Given the symptoms are relatively vague and slowly progressive, the disease may first be misdiagnosed as asthma or bronchiolitis, and the true diagnosis and treatment are often delayed.<sup>3,9-11</sup> Although women are commonly diagnosed just before or around the time of menopause, some manifest the disease earlier in their childbearing years. While no concrete figures are reported in the current literature, up to 5% of patients in the North American Airway Collaborative iSGS1000 cohort reported symptoms which first began in pregnancy.<sup>12</sup> Scant case reports in obstetrics and gynecology literature further demonstrate the uncommon occurrence of iSGS arising during pregnancy.<sup>13-15</sup> Our survey study is

the first to specifically provide important preliminary impressions of the iSGS disease process during pregnancy from the perspective of the patient.

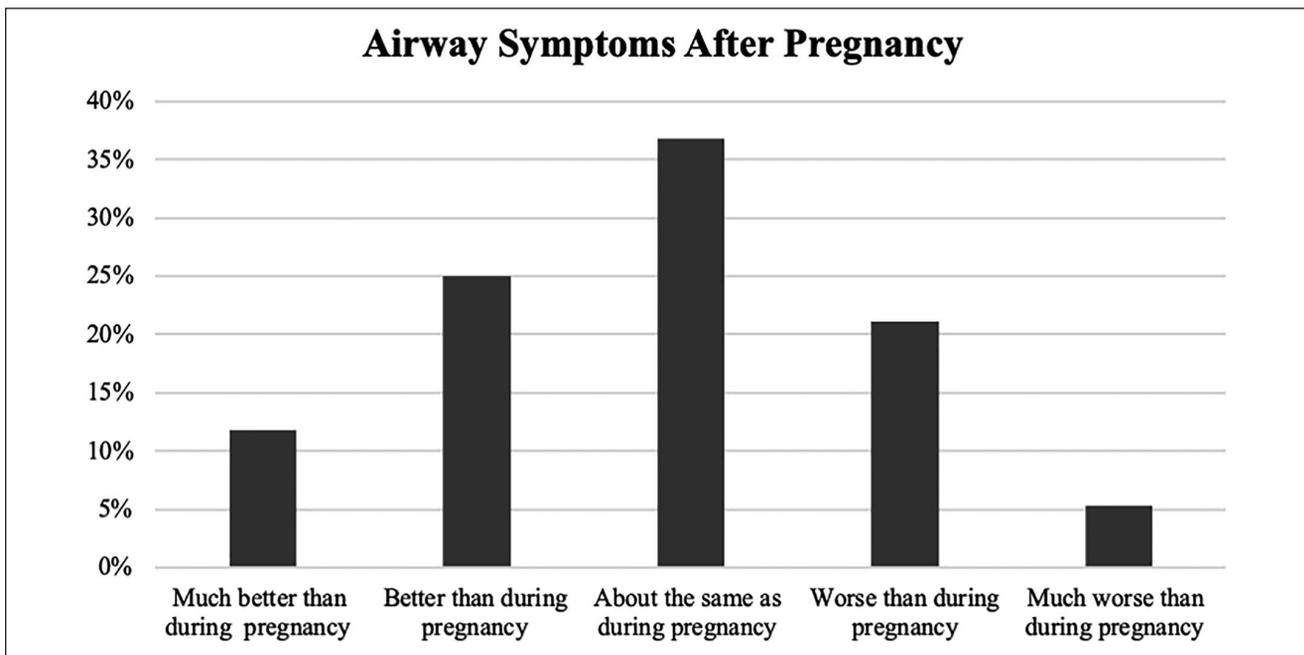
Over 400 women with iSGS responded to our survey, with approximately 84.7% of respondents reported receiving a diagnosis of iSGS prior to menopause during child bearing years. Our survey data shows approximately 5.8% of women received their diagnosis of iSGS in the context of pregnancy. No definitive cause of iSGS has been identified, but because the initial presentation often coincides with menopause, the underlying belief is that the disease process is hormonally mediated.<sup>16-18</sup> Some evidence suggests that subglottic narrowing and deposition of collagen is mediated by estrogen.<sup>19,20</sup> Certainly, hormonal changes occur during pregnancy as well, thus, presentation during pregnancy would not be unexpected. Our results raise the question of whether more women are susceptible to development of peripartum iSGS than previously suspected. Interestingly, patients in this survey did not indicate that there was worsening of their airway symptoms with use of oral contraceptives or post-menopausal hormones.

Only 25.5% of respondents had ever been pregnant while carrying a diagnosis of iSGS. However, of patients who dealt with iSGS during pregnancy, 71.1% reported a significant increase in the severity of their airway symptoms. Of note, this worsening of symptoms did not correspond to an increase in the number of either operative nor in-office procedures during pregnancy. However, slightly more operative procedures were completed after pregnancy. Of note, among patients that underwent operative management during pregnancy, approximately 61.5% of these patients again underwent operative management of their disease in the months after delivery. This suggests that there may be a more recalcitrant group of iSGS patients who may be more adversely affected in the months during and after pregnancy. More research is needed to determine the etiology of this increased percentage of postpartum surgery; however, one could surmise that some patients postponed their procedures until after delivery. Patients, although uncomfortably symptomatic, may have elected to delay procedures in order to protect their babies from potentially harmful side effects associated with general anesthesia.

Although iSGS was historically managed with operative endoscopic resection and dilation, our practice has shifted to more in-office management with growing familiarity of the disease process and better equipment.<sup>21-23</sup> In emergent cases, sometimes a tracheostomy has been performed and used throughout gestation and was then followed up by cricotracheal resection or endoscopic resection with eventual tracheostomy decannulation.<sup>14,24</sup> Ultimately, communication is key between the head and neck surgery team and obstetrics when considering any surgical intervention. The American College of Obstetricians and Gynecologists have established guidelines on nonobstetric surgery during



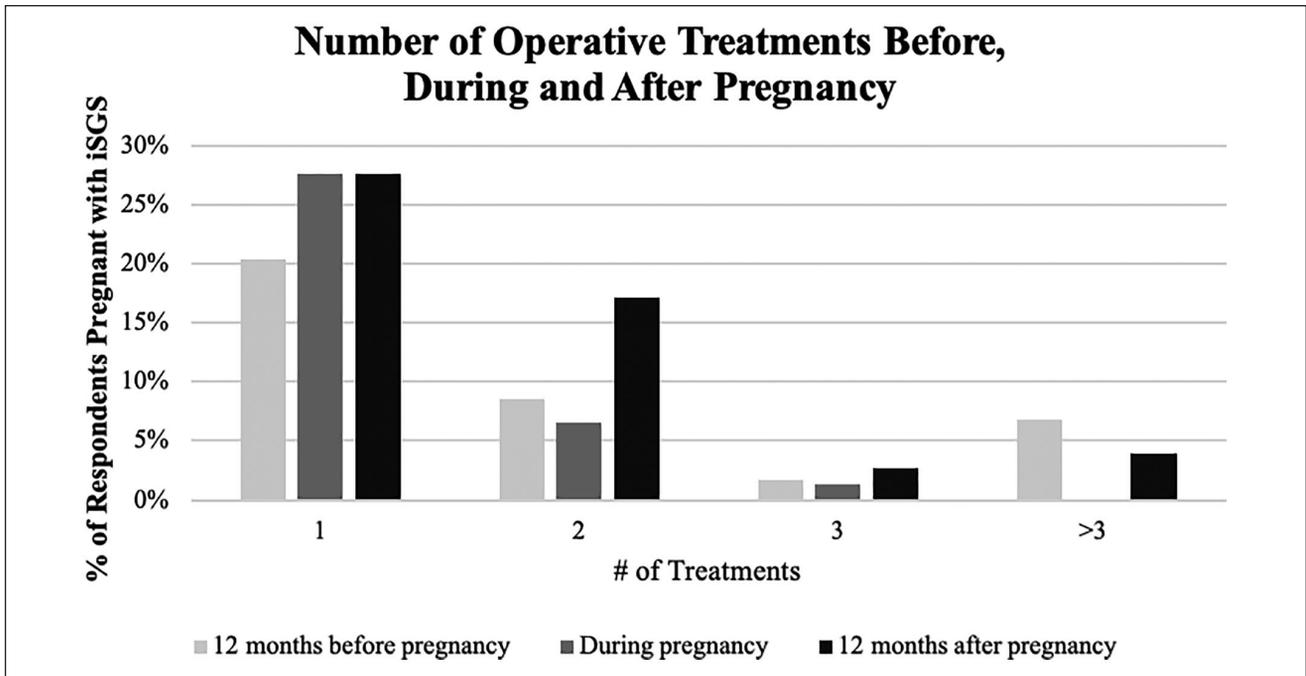
**Figure 1.** Frequency of airway symptoms among survey participants during pregnancy.



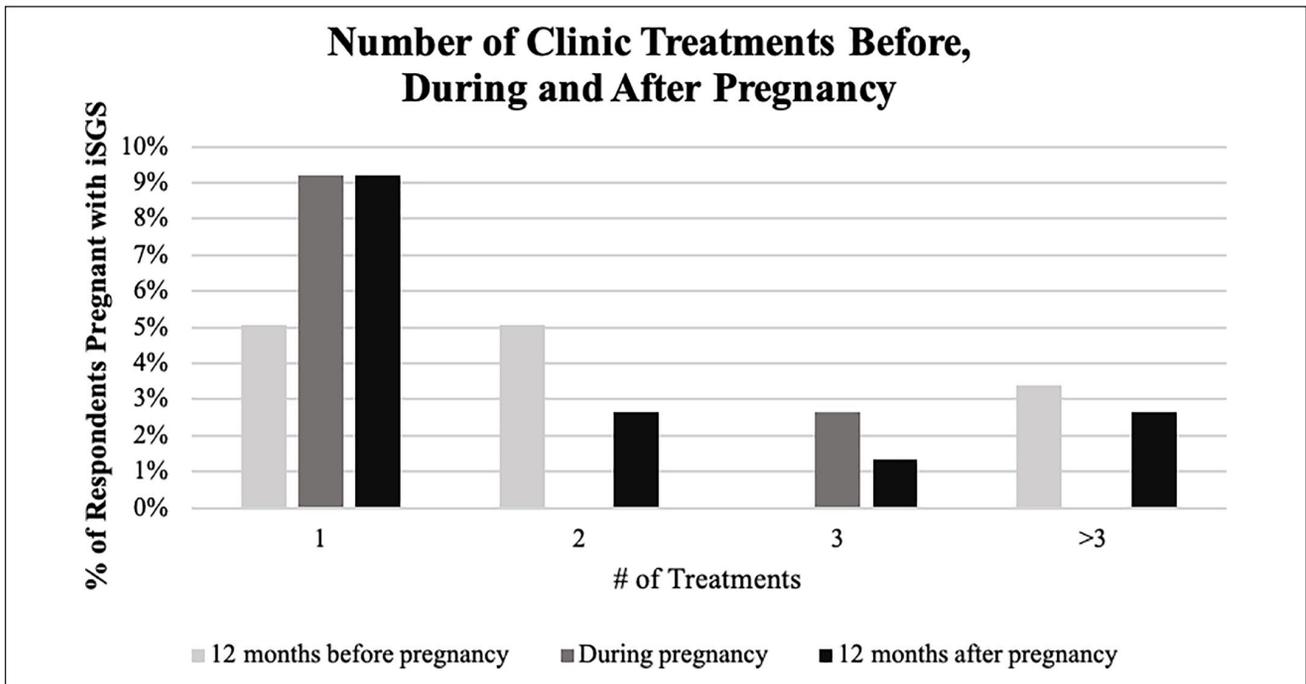
**Figure 2.** Frequency of airway symptoms among survey participants after pregnancy.

pregnancy, with these recommendations suggesting that surgery should not be denied to a pregnant woman if it is medically necessary, regardless of which trimester they are in.<sup>25</sup> While there is a risk of preterm labor, the risks associated with delaying surgery can also contribute to poor perinatal outcomes, which could be considered to be more

dangerous than the surgery itself.<sup>26</sup> Our practice has generally moved to more clinic-based procedures for management of iSGS during pregnancy, however, the decision for surgery should include all key stakeholders, including the patient, the head and neck surgery team, the patient's obstetrician, and anesthesia.



**Figure 3.** Frequency of operative treatments 12months before pregnancy, during pregnancy and 12months after pregnancy among survey participants.



**Figure 4.** Frequency of clinic treatments 12 months before pregnancy, during pregnancy and 12 months after pregnancy among survey participants.

Unfortunately, 40.8% of patients who dealt with iSGS during pregnancy felt this adversely affected their decision to have children in the future. While the survey question did not probe further, this may be due to difficulties during and

shortly after pregnancy. Just over half of patients with iSGS experienced pregnancy complications with approximately 21.7% of them suffering from preeclampsia. At least one case report diagnosed iSGS in the context of urgent

intubation for a pregnant patient with preeclampsia.<sup>13</sup> One study notes significantly narrower airways in preeclamptic patients as compared to uncomplicated pregnant and non-pregnant patients.<sup>27</sup> Another found increased soft tissue thickness at the hyoid bone in patients with preeclampsia compared to normal pregnancy, but there were no differences at the level of the glottis and the subglottis was not evaluated.<sup>28</sup> The correlation and importance of these findings remain unclear, and further research is needed to explore possible connections regarding preeclampsia in pregnant iSGS patients and if iSGS has compounding effects on airway caliber.

Although we hoped to capture an accurate view of the patient experience of iSGS during pregnancy by polling a large, international iSGS support group, our findings may be affected by the inherent nature of a survey study. Participation bias and recall bias are certainly possible. It is important to note that this study was also completely anonymous without specific enrollment criteria, other than self-reporting that the participant was a patient with idiopathic subglottic stenosis. Importantly, Tanner et al. used similar methodology as ours to evaluate nebulizer use among subglottic stenosis patients enrolled in the same Facebook group.<sup>29</sup> Furthermore, question stems and answer choices were sensitively crafted and thoroughly reviewed prior to distribution; however, misinterpretation of questions and answer choices are possible. We feel that this study provides important insight into the patient experience of pregnancy and iSGS. Given the increasing symptom severity during pregnancy, otolaryngologists should consider the implementation of non-invasive clinic procedures such as in-office subglottic steroid injection in the treatment of pregnant women with iSGS.<sup>22</sup> Future avenues for research include further evaluating the interplay between hormonal changes and iSGS. This may provide increased clarity on how pregnancy affects presentation and severity of iSGS.

## Conclusions

We present a preliminary survey-based analysis of the patient experience of iSGS during pregnancy. Subjective worsening in airway symptoms was noted in up to 71.1% of participants, however, this did not lead to more operative or office-based procedures. The majority of patients with iSGS felt that the disease adversely affected future pregnancy decisions. Further research is needed to characterize the disease process and management options for this unique and previously under-recognized group within the iSGS population.

## Conference Presentation

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\*Presented as E-poster given COVID-19.

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## Supplemental Material

Supplemental material for this article is available online.

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