TITLE: PATIENT REPORTED DATA OF DESMOID TUMORS DURING AND AFTER PREGNANCY FROM AN INTERNATIONAL NATURAL HISTORY STUDY

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OBJECTIVES: Desmoid tumors (DTs), a subtype of sarcoma, are a rare disease with variable and unpredictable clinical course. It has an incidence of 5-6 per million/year with a median age of 30-40, affecting females more than males. To date, there are very few studies that provide data on the impact of pregnancy to DT behavior. Patient reported outcomes of pregnancy with a DT are described here.

METHODS: The web-based natural history study launched September 2017 in collaboration with the National Organization of Rare Disorders. It contains 15 surveys covering diagnostics, disease, treatment, care management, and quality of life. The pregnancy survey was conditionally provided to participants who reported their sex as female and age > 18 years old. The questionnaire poses questions about the status of the participant's DT(s) before, during, and following pregnancy.

RESULTS: Of the 696 participants that have consented and started all surveys, 182 have completed the pregnancy survey. Eighteen of the 182 (9.9%) reported that they had a DT at the time of becoming pregnant. Thirteen are from the United States, and one from each of Australia, United Kingdom, Jersey, China, and Belgium. Of these 18 pregnancies, five (27.7%) were premature (less than 37 weeks) and 3 of those did not result in live birth (16.7%). Comparatively, in the United States, 10.3% of all pregnancies end in premature deliveries and 10-20% of miscarriages are reported.

During pregnancy, the participants reported that their DT grew (n=7, 41%), shrank (n=2, 12%), and was stable (n=4, 24%). For most women, the DT behaved the same after pregnancy as it did during pregnancy. For one participant, the DT was stable during pregnancy and grew postpartum, while another participant had DT growth during pregnancy and stable DT postpartum. Treatment and surveillance data is not currently collected during pregnancy. The distribution of the location of the DT for the pregnancy survey was similar to that of the entire dataset ($p = 0.48$). There were lower numbers of head and neck DTs (0% vs. 10%) and chest wall (5% vs. 18%) in the pregnancy dataset (0% vs. 10%), but higher rates of abdominal wall DTs (25% vs. 18%). Six participants reported having FAP mutations, and two reported *CTNNB1* mutations. There did not appear to be a correlation between mutations and preterm or live births.

CONCLUSION: DTs are most common in females with a median age of 30-40 years. However, few studies have investigated the effects of pregnancy on DTs. The pregnancy dataset from the Natural

History Study indicates that DTs do not change behavior due to pregnancy and presumably exposure to hormone fluxuations. In addition, DT location and mutation status do not seem to be correlated with preterm or live births. In the future, the study will capture the use of active surveillance during pregnancy.

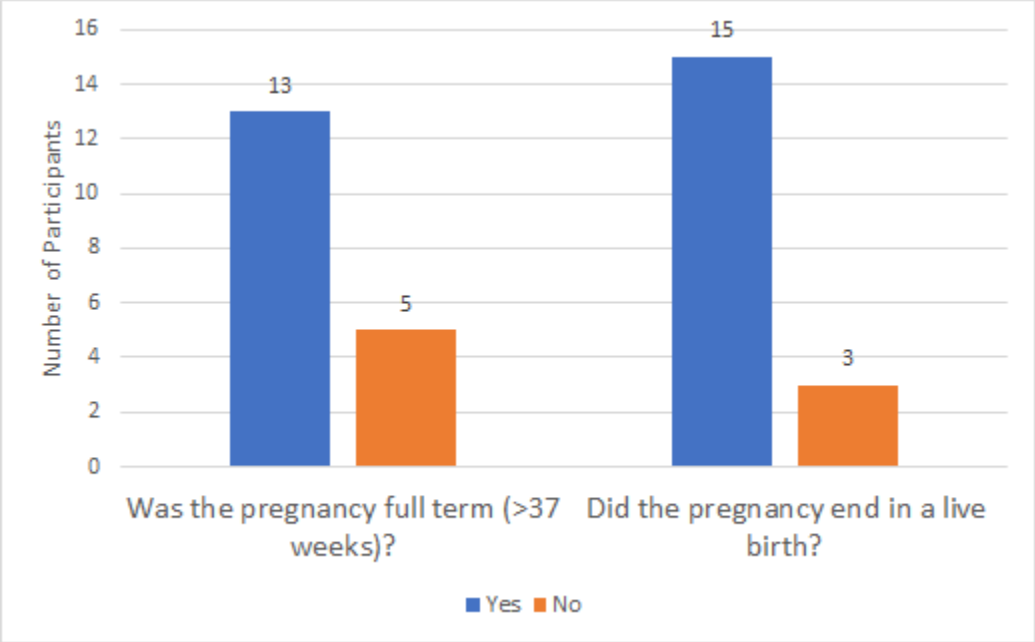

Figure 1. The participants who reported if their pregnancy was full term (yes, no) and if their pregnancy resulted in a live birth (yes, no). While there is a slightly higher rate of a pre-term delivery with a DT, the rate of pregnancy loss does not appear to be dissimilar to rates found in a general population.

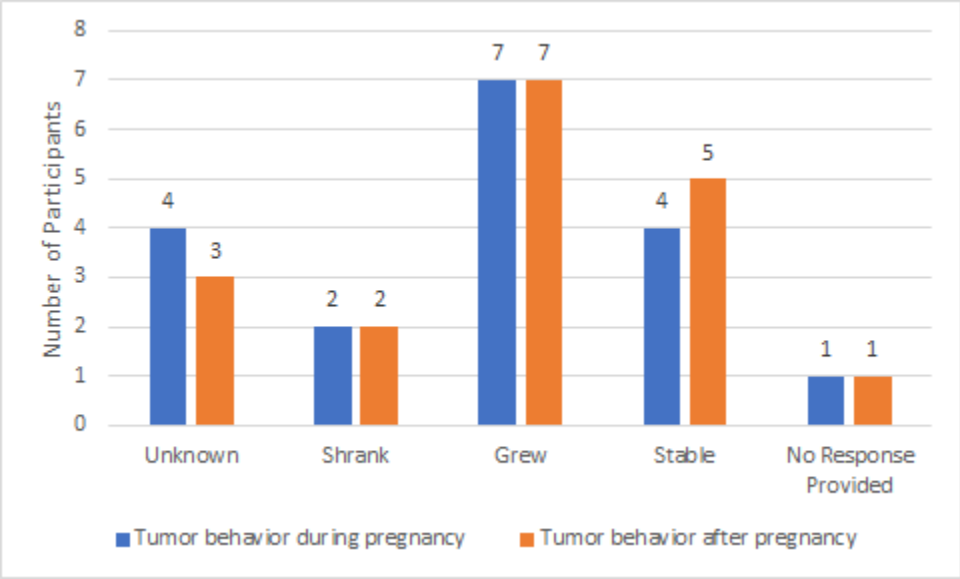

Figure 2. Characteristics of how the DT behaved during the pregnancy and after delivery. With the small number of respondents thus far, no trends are apparent with the impact of pregnancy hormones on the growth of the DTs during or after pregnancy. No response reflects that the participants did not answer that question.