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

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THE DESMOID TUMOR RESEARCH FOUNDATION

1. BACKGROUND AND AIMS

Desmoid tumors (DTs), a subtype of sarcoma, are a rare disease with variable and unpredictable clinical course. There is an incidence of 5-6 individuals diagnosed per million/year with a median age of 30-40, affecting more females than males.

To date, there are very few studies that provide data on the impact of pregnancy to DT behavior[1,2]. Patient reported outcomes of pregnancy with a DT are described here. The objectives of which include sharing the data from self-reported cases of pregnancy with DT and describe the patient experience to inform the community.

2. METHODS

The DTRF Patient Registry is designed to collect data for a prospective longitudinal web-based observational Natural History Study (NHS). Participants with DTs will be followed throughout the course of their lives with either the participant or authorized respondents contributing data at varying intervals throughout the course of the study.

The pregnancy survey was conditionally provided to participants who reported their sex as female. The questionnaire poses questions about the status of the participant's DT(s) before, during, and following pregnancy. The conditions of the survey were modified after the study initiation to only collect information for participants > 18 years old.

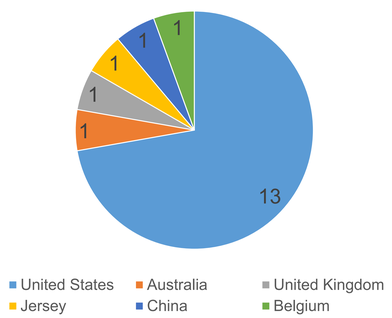
The DTRF launched the patient registry and NHS in 2017 and was one of the inaugural organizations in the I AM RARE project. This registry and study has ethical approval for global data collection from North Star Review Board IRB (Protocol Number: NB100030).

Data was downloaded for analysis on June 6, 2021.

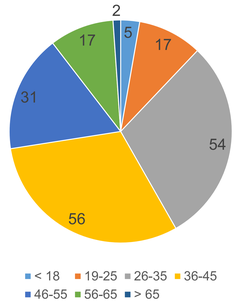
3. FEMALE PARTICIPANTS

There were 182 females who have completed the pregnancy survey. While most of the participants who have completed this survey are in the United States (156/182), there are a handful of participants in the EU and Asia. Because of platform limitation, the study is limited to English language.

Eighteen of the 182 (9.9%) reported that they had DT at the time of having their first pregnancy. Thirteen of the 18 pregnant with DT are from the United States, and one from each of Australia, United Kingdom, Jersey, China, and Belgium. Of note: an additional 39 patients responded unknown if DT was present when they became pregnant (not depicted).



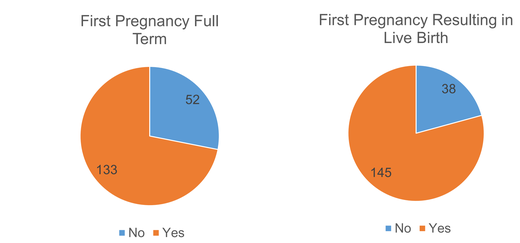
4. AGE OF FEMALE PARTICIPANTS



Of the 182 females who completed the survey, the majority were between 26-35 (54/182) and 36-45 (56/182) years at diagnosis.

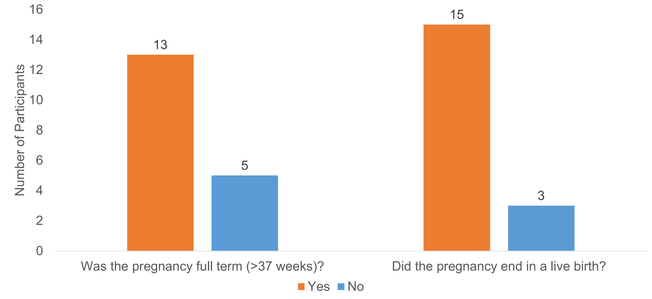
There were 5 participants noted to be under the age of 18. These surveys were completed by legally authorized representatives and did not report pregnancies.

5. ALL PREGNANCY SURVEY OUTCOMES



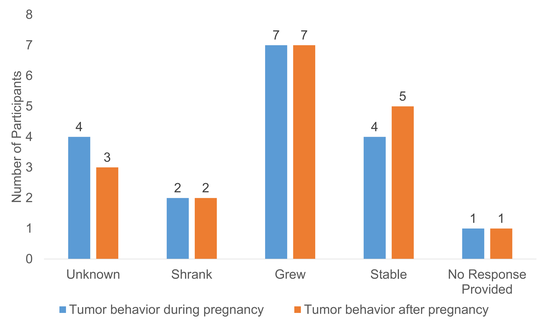
For those not having a DT during the first pregnancy, reported outcome: Full Term was 71.9% (N=185*) and the reported outcome: Live Birth was 79.2% (N=183*).

There are similar pregnancy outcomes between those participants with a known DT present during pregnancy as compared to those DT patients that completed the pregnancy outcomes survey. These were not tested for significance given the small number of participants.



Of the 18 pregnancies reported, five (27.8%) were preterm (less than 37 weeks) and 3 of those did not result in live birth (16.7%). Comparatively, in the United States, 9.54% of live births are preterm deliveries and 10-20% of miscarriages are reported[3].

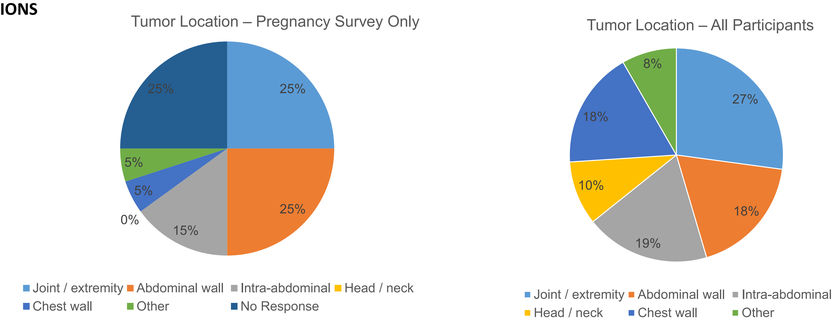
6. TUMOR BEHAVIOR DURING AND AFTER PREGNANCY



During pregnancy, the participants reported that their DT grew (n=7, 41%), shrank (n=2, 12%), or 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.

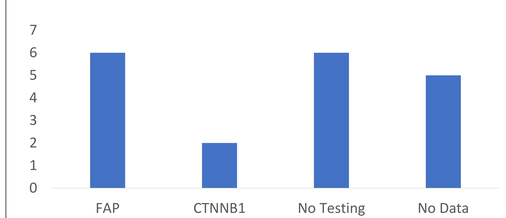
Treatment and surveillance data is not currently collected during pregnancy.

7. TUMOR LOCATIONS



The distribution of the location of the DT for the pregnancy survey was comparative that of the entire dataset (p = 0.48). There were fewer numbers of head and neck DTs (0% vs. 10%) and chest wall (5% vs. 18%) in the pregnancy survey only dataset, but higher rates of abdominal wall DTs (25% vs. 18%).

8. MOLECULAR RESULTS FOR PREGNANT PARTICIPANTS



Six participants reported having mutations relating to FAP (Familial adenomatous polyposis), and two reported *CTNNB1* mutations. Additionally, six participants reported not being tested and no data.

There did not appear to be a correlation between mutations and preterm or live births (not shown).

9. SUMMARY AND FUTURE DIRECTIONS

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 data from this NHS suggests that DTs do not change behavior during or due to pregnancy and the presumable exposure to hormone fluctuations. In addition, DT location and mutation status do not seem to be correlated with pregnancy outcomes like preterm or live births.

The NHS committee anticipated and planned for on-going study modifications as the data matures. Specific to this abstract, there is an opportunity to modify the surveys to collect additional details related to treatment status while pregnant and to include an active surveillance option during pregnancy. Also as mentioned, 39 participants responded unknown if DT was present during pregnancy; modifications to the survey will collect additional clarifying information. These edits will help strengthen the data from the NHS and better inform the community.

The DTRF NHS researchers are extremely grateful to the participants, caregivers, and the NORD staff for their ongoing engagement in this important study.

References: 1. Ann Surg. 2014 May;259(5):973-8. 2. Cancers (Basel). 2012 Mar; 4(1): 184–192. 3. Morb Mortal Wkly Rep 2016;65:1181–1184