

TITLE: Quality of Life and Tumor Location in Patients with Desmoid Tumors: Data from the Desmoid Tumor Research Foundation Natural History Study

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ABSTRACT KEY WORDS: Desmoid tumors, Natural History Study, Patient Registry

ABSTRACT OBJECTIVE: The Desmoid Tumor Research Foundation launched the DTRF patient registry in partnership with the National Organization for Rare Disorders. The registry launched September 2017 and contains surveys diagnostics, treatments, disease monitoring, and quality of life. The early demographics and quality of life measures are described within.

BACKGROUND AND OBJECTIVES: Desmoid tumors occur in connective tissues of the body. These noncancerous tumors may invade surrounding tissues resulting in compromised organ function and complications. As few as 5 per 1 million people are diagnosed with desmoid tumors annually, which may be an underestimate of the actual affected population due to difficulty in correctly diagnosing the disease. To improve awareness of desmoid tumors and better inform treatment development, DTRF in partnership with the National Organization for Rare Disorders launched the DTRF patient registry. Here, we describe patient demographics, tumor location, and QOL in registry patients.

METHODS: The registry launched September 2017 and contains 15 surveys covering diagnostics, disease, treatment, care management, and quality of life. As of January 2019, 357 patients have completed 2,371 surveys.

RESULTS: Registry participants are mostly white (88%, 313/357), female (81%, 277/343), and reside in 27 countries with 80% (285/357) US-based. Median age at diagnosis is 33 and the time from onset of symptoms to diagnosis was more than 1 year for 54% (189/352) of respondents. Desmoid tumor location was reported for 119 respondents with tumors at time of data collection. Most prevalent tumor locations were joint /extremities (39%, 47/119), intra-abdominal (24%, 28/119), and chest wall (24%, 29/119). Multiple tumor locations were indicated for 22% (26/119) of patients. QOL reported as very good or excellent ranged from 28% to 60% of patients, depending on tumor location.

CONCLUSION: Patients with desmoid tumors have varied QOL and tumor locations. Data collection through the registry is ongoing.