

# QUALITY OF LIFE (QOL) AND TUMOR LOCATION IN PATIENTS WITH DESMOID TUMORS; DATA FROM THE DESMOID TUMOR RESEARCH FOUNDATION (DTRF) NATURAL HISTORY STUDY

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THE DESMOID TUMOR RESEARCH FOUNDATION

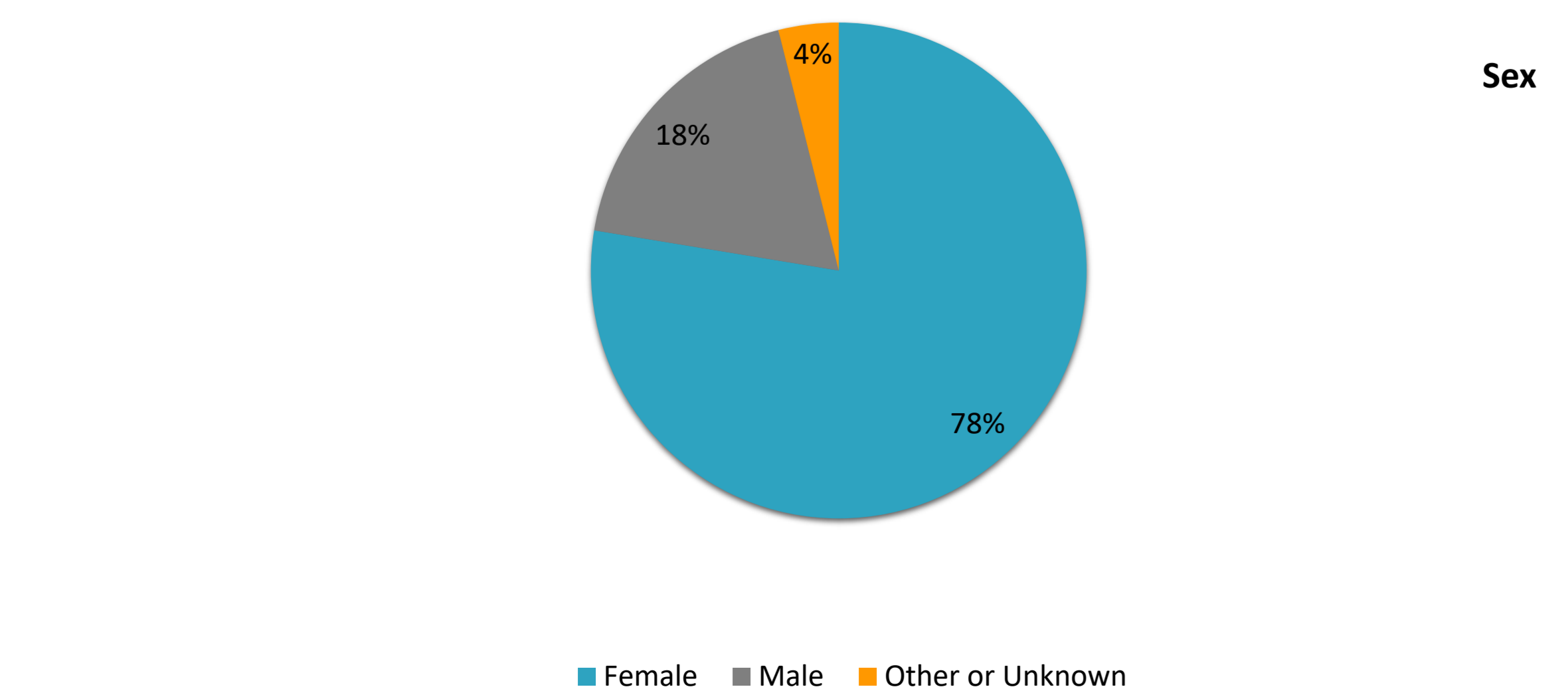
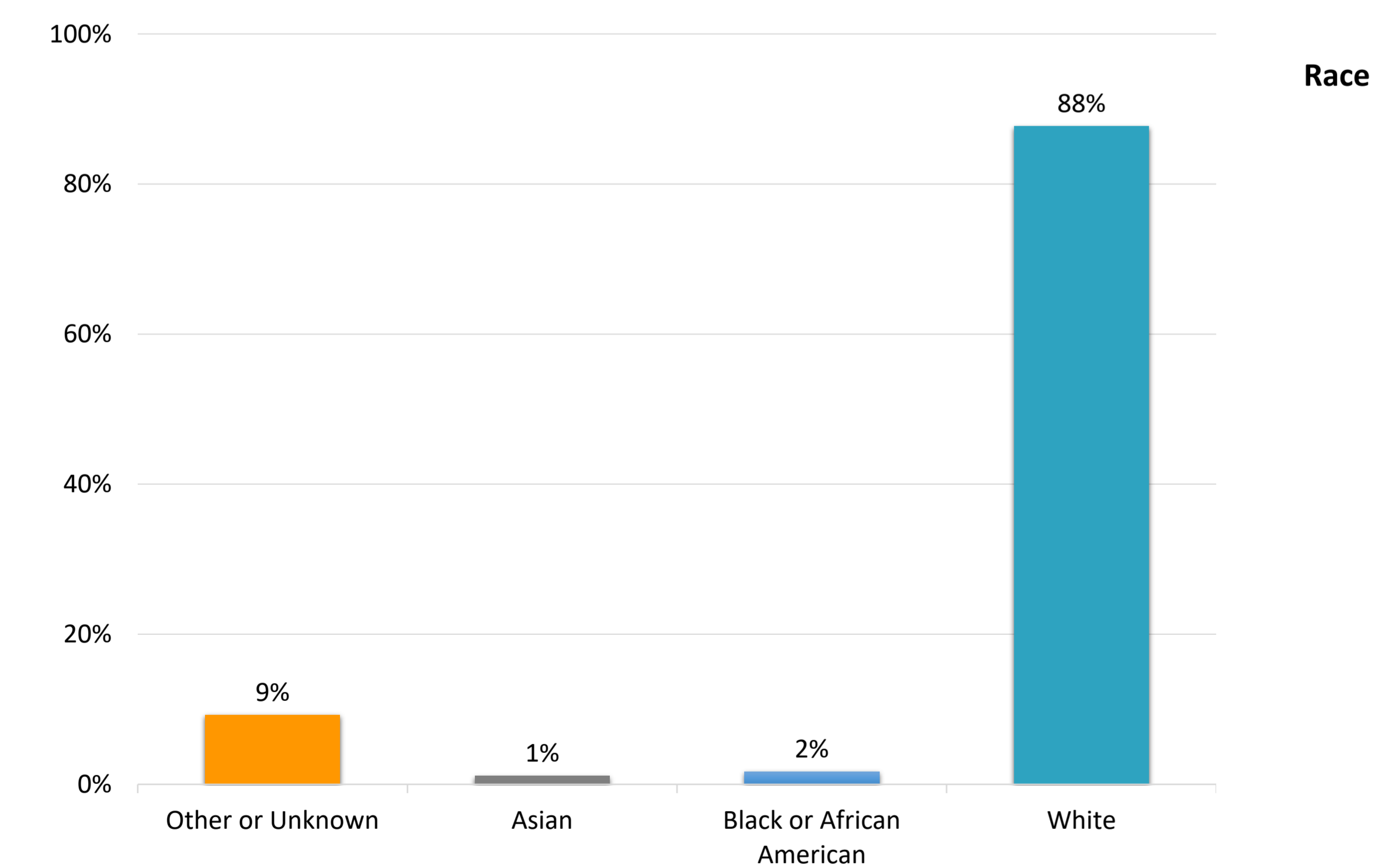
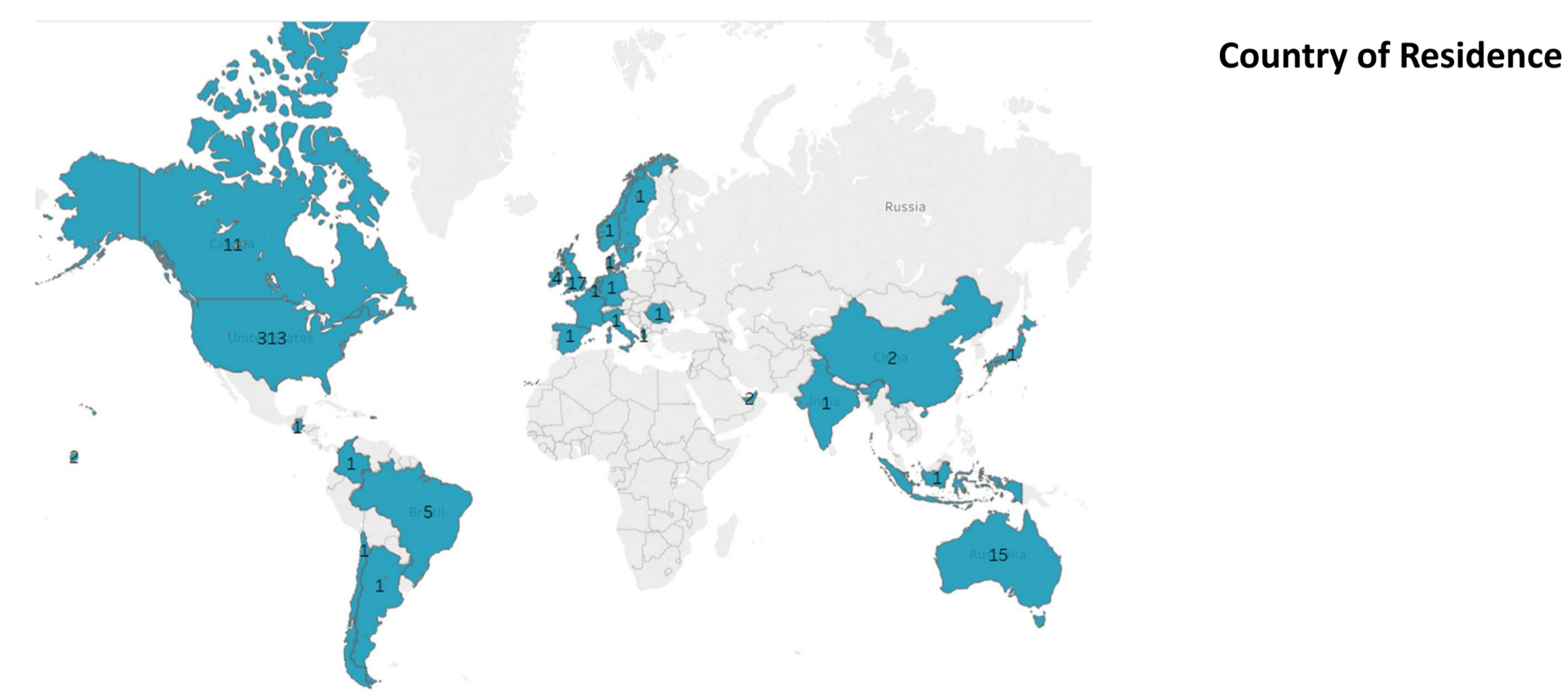


## 1. BACKGROUND AND AIMS

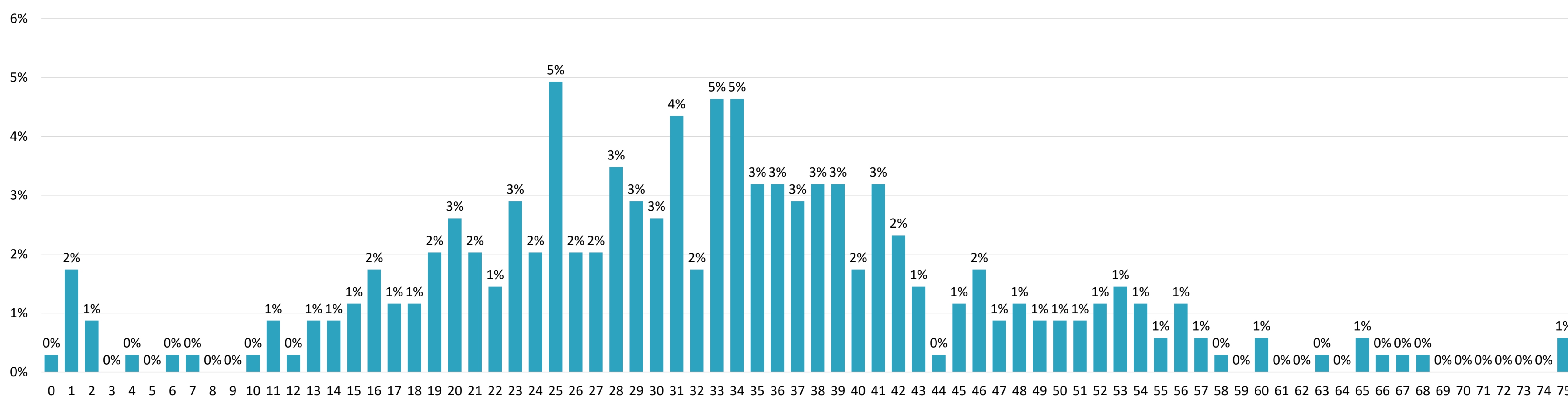
Desmoid tumors occur in connective tissues of the body. These noncancerous tumors may invade surrounding tissues resulting in compromised organ function and complications. As few as 5-6 per 1 million people are diagnosed with desmoid tumors annually, which may be an underestimate of the actual affected 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 natural history study. Here, we describe patient demographics, tumor location, and QOL in registry patients.

## 2. METHODS

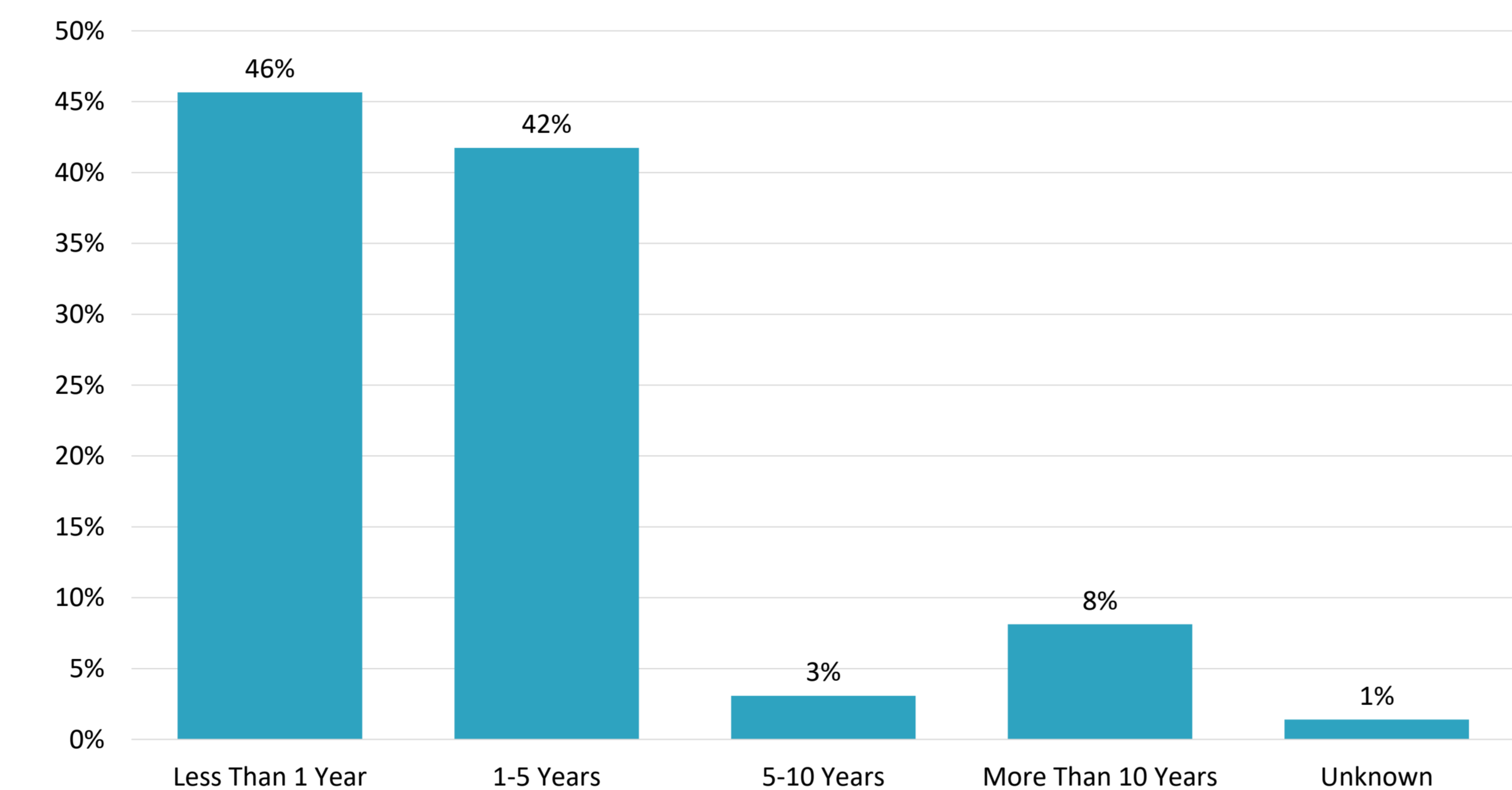
The registry initiated February 2017 and contains 15 surveys covering diagnostics, disease, treatment, care management, and quality of life. As of January 2019, 357 patients have completed 2,371 surveys.



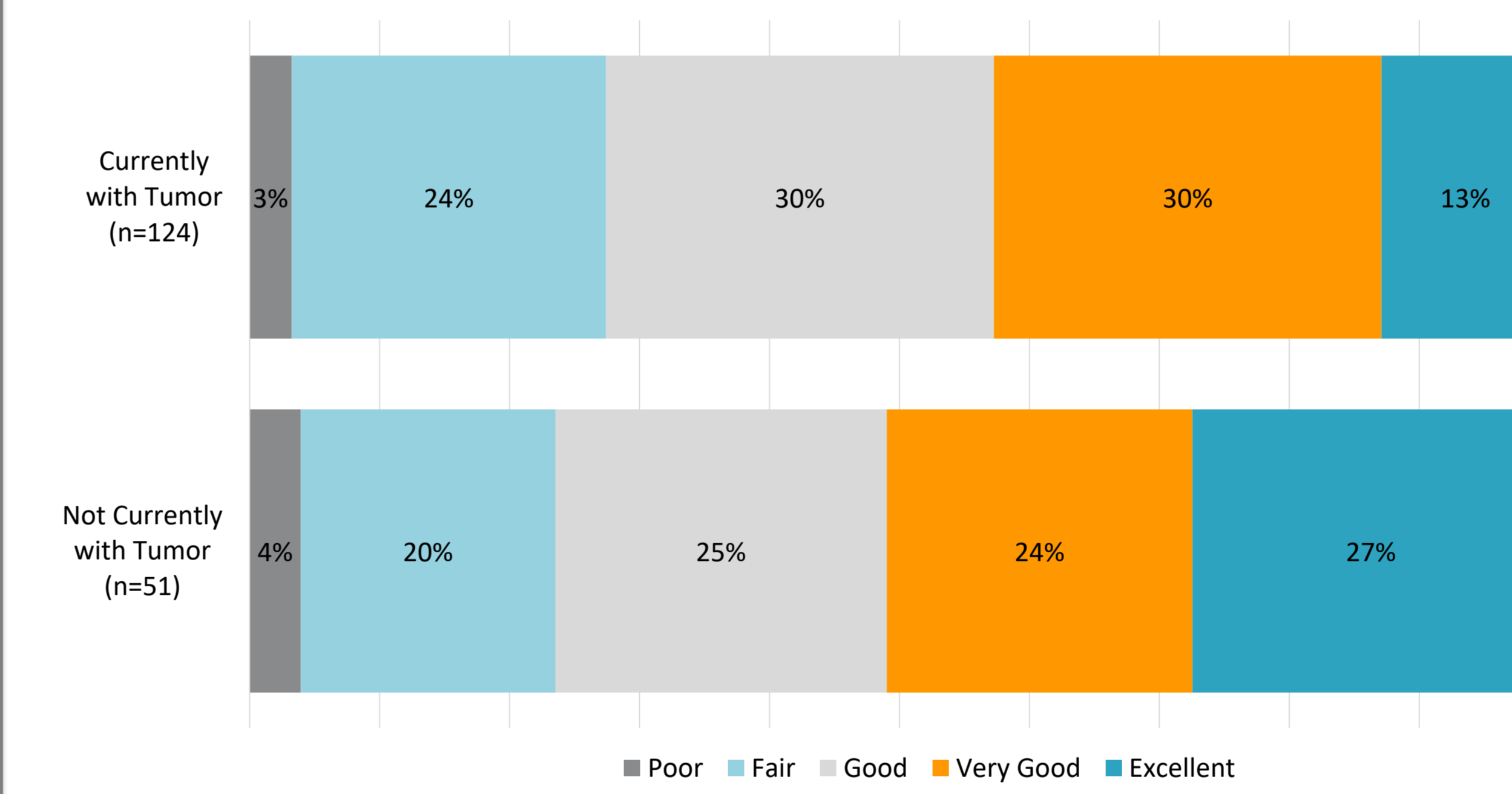
## 3. AGE (YEARS) AT DIAGNOSIS (N=345)



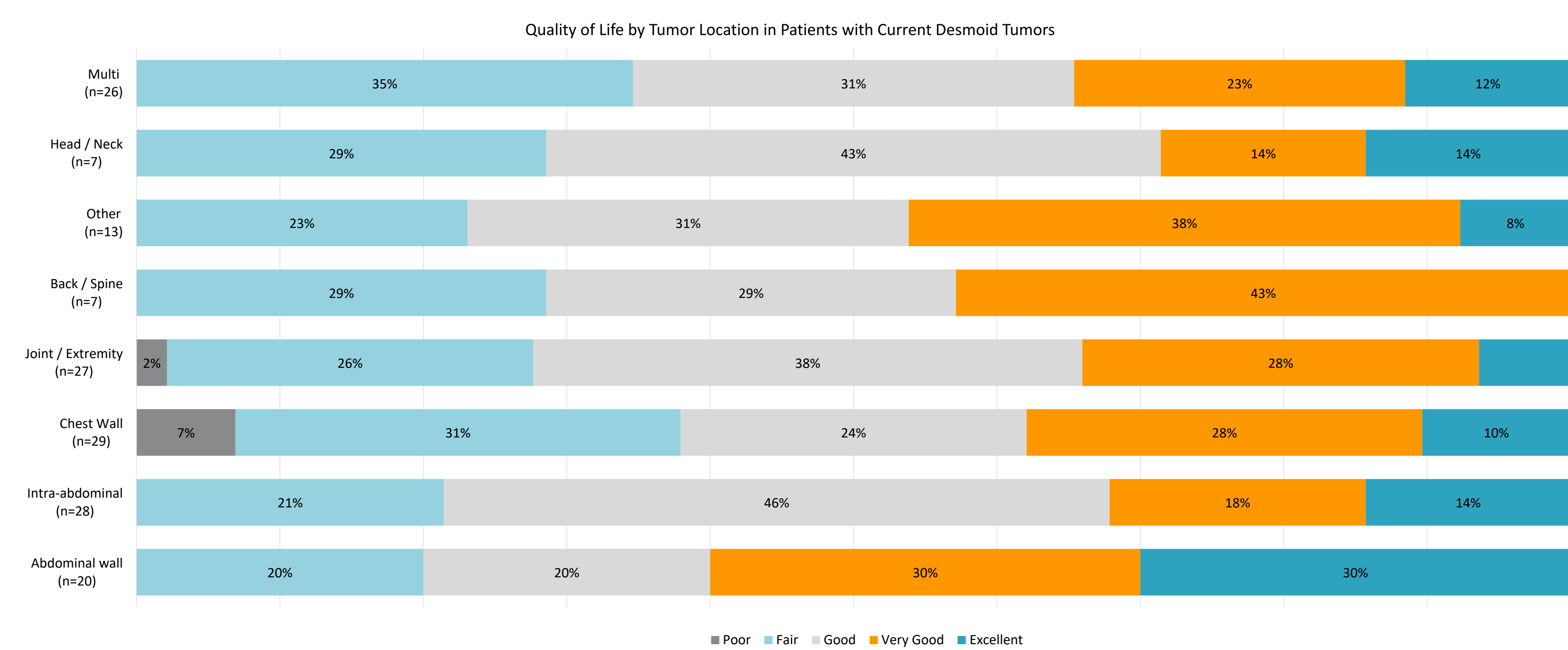
## 4. TIME FROM ONSET OF SYMPTOMS TO DIAGNOSIS (N=357)



## 5. QUALITY OF LIFE BY TUMOR STATUS

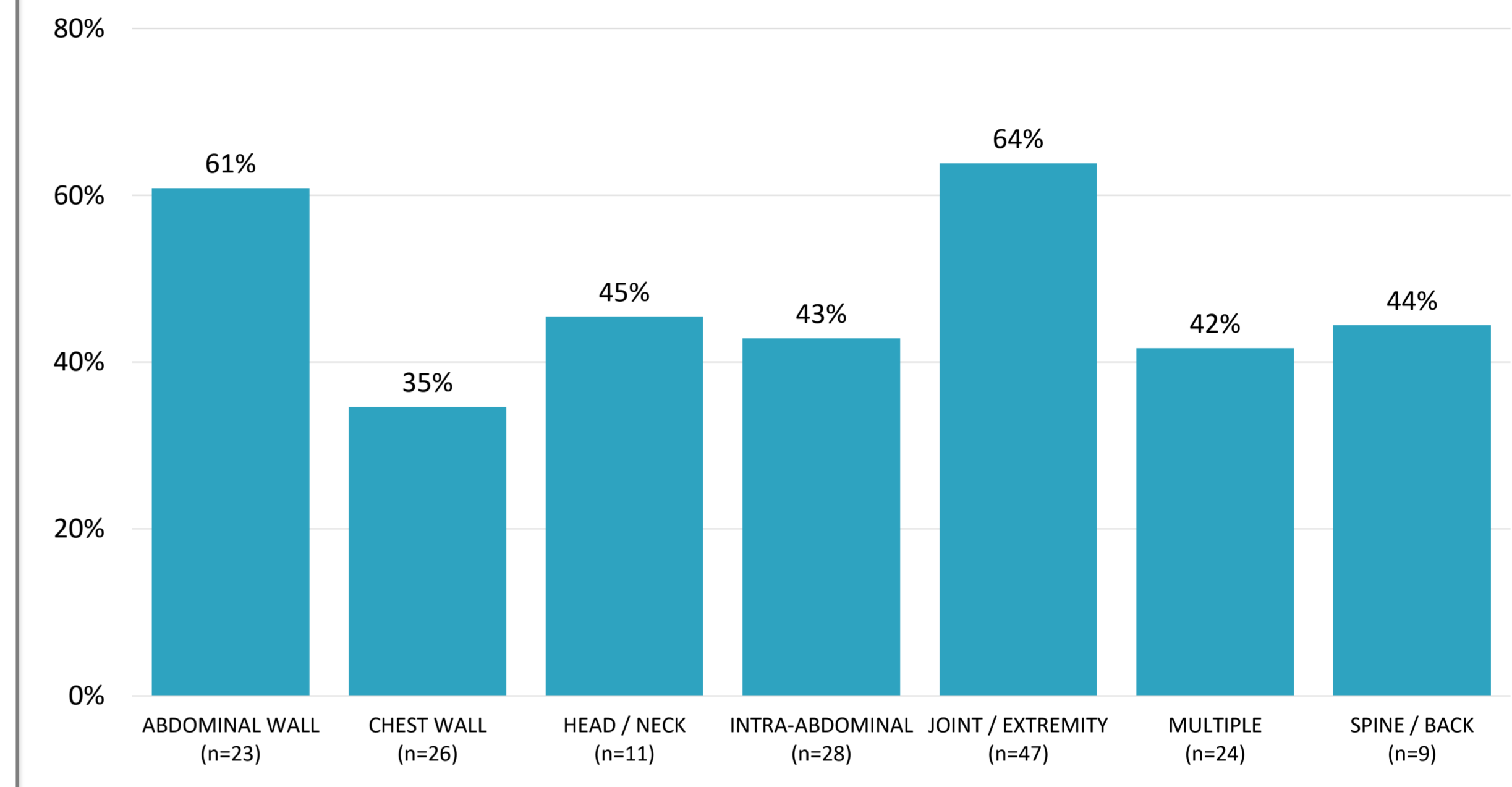


## 6. QUALITY OF LIFE BY LOCATION OF TUMOR



## 7. MISDIAGNOSIS BY INITIAL TUMOR LOCATION

Misdiagnosis rates varied by initial tumor location and are highest for abdominal wall and joint / extremity. Assessments of proportions using z-test with Bonferroni correction identified only joint / extremity as more prevalent (e.g. reaching significant of  $p \leq 0.05$ ) in misdiagnosed patients.



## 8. SUMMARY

Patients with desmoid tumors have varied QOL and tumor locations. Data collection through the registry is ongoing and essential to raising awareness of this disease and improving care for these patients.

### Location

- Registry participants are located in 28 countries
- Participants are mostly US-based (80%, 313/390)

### Race and Sex

- Registry participants are mostly white (88%, 313/357) and female (78%, 277/357)

### Age at Diagnosis

- The median age of diagnosis is 33 years

### Time from Onset of Symptoms to Diagnosis

- 46% of participants were diagnosed within one year after onset of symptoms
- Most participants (88%) were diagnosed within 5 years of onset of symptoms

### Quality of Life by Tumor Status

- The number of participants who rated their quality of life as excellent varied significantly depending on whether or not the participant currently had a tumor at the time ( $z=2.320$ ,  $p=0.020$ , significant at  $p < .05$ ).

### Quality of Life and Misdiagnosis by Location of Tumor

- 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.
- In this study of patient registry data, nearly half (49% (84/171) of respondents indicated initial misdiagnosis.
- The challenge of correct initial diagnosis remains. Data collection through the DTRF patient registry continues with increasing awareness of desmoid tumors.

*Study findings may not be generalizable to all individuals in the desmoid tumor community. These analyses represent data from the sample of individuals who participated in the Desmoid Tumor Patient Registry.*