

Supplementary Material

Desmoid Tumors: A Comprehensive Review

Authors: Meryem Bektas, PhD, MBA^a (0000-0001-8080-7557); Timothy Bell, MHA^b (0000-0001-8909-5293); Shahnaz Khan, MPH^a (0000-0002-5075-2732); Brad Tumminello, PharmD^b (0009-0004-0967-7330); Maria M Fernandez, PhD, MBA^a (0000-0002-7361-333X); Catriona Heyes, MSc^a; Ana B. Oton, MD^b (0000-0002-6126-7353)

^aRTI Health Solutions, Research Triangle Park, NC, USA

^bSpringWorks Therapeutics, Inc., Stamford, CT, USA

Address for correspondence:

Timothy Bell

SpringWorks Therapeutics, Inc.

5310 S. Alston Avenue

Durham, NC, 27713, USA

Tel. 1 919-698-3583

email: Timothy.Bell@springworkstx.com

METHODS

Search Strategies

Tables S1-S3 present the search strategies developed for PubMed, Embase and Cochrane database, respectively, to identify studies included in this manuscript. Table S4 presents the Embase search strategy to identify conference abstracts published in the last 5 years up to 30 November 2021 (original search date). Abstracts presented to conferences in 2022 up to the submission of this manuscript were also reviewed.

Table S1. PubMed Literature Search Strategy: Burden of Illness of DT

Limits: last 10 years; English only; humans; no comments, letters, editorials, or case reports

Search number	Search terms
Disease	
#1	"fibromatosis, aggressive"[Majr] OR ("fibromatosis"[Title] AND "aggressive"[Title]) OR "aggressive fibromatosis"[Title] OR "desmoid type fibromatosis"[Title] OR ("fibromatosis"[Title] AND "desmoid"[Title]) OR "desmoid tumor"[title] OR "desmoid tumours"[Title] OR "desmoid tumors"[Title] OR "desmoid tumour"[Title] OR "desmoid fibromatosis"[Title] OR "deep musculoaponeurotic fibromatosis"[Title] OR "musculoaponeurotic fibromatosis"[Title]
Epidemiology	
#2	#1 AND ("Fibromatosis, Aggressive/epidemiology"[Majr] OR "Epidemiology"[Majr] OR "Morbidity"[Majr] OR "Incidence"[Majr] OR "Prevalence"[Majr] OR "Risk Factors"[Majr] OR epidemiolog*[Title] OR incidence*[Title] OR prevalence[Title] OR morbidit*[Title] OR risk*[Title])
Humanistic burden	
#3	#1 AND ("Quality of Life"[Mesh] OR "Self Report"[Mesh] OR "Activities of Daily Living"[Mesh] OR "Patient Satisfaction"[Mesh] OR "Patient Preference"[Mesh] OR "Quality-Adjusted Life Years"[Mesh] OR "Health Status"[Mesh] OR "Stress, Psychological"[Mesh] OR burden[Title/Abstract] OR "quality of life"[Title/Abstract] OR "life quality"[Title/Abstract] OR "QoL"[Title/Abstract] OR "hrql"[Title/Abstract] OR "hrqol"[Title/Abstract] OR "EuroQoL"[Title/Abstract] OR "EQ5D"[Title/Abstract] OR "EQ5D"[Title/Abstract] OR "quality of well being"[Title/Abstract] OR "well being"[Title/Abstract] OR "SF-6D"[Title/Abstract] OR patient report*[Title/Abstract] OR proxy report*[Title/Abstract] OR self report*[Title/Abstract] OR questionnaire*[Title/Abstract] OR "functional status"[Title/Abstract] OR "activities of daily living"[Title/Abstract] OR "physical function"[Title/Abstract] OR "patient satisfaction"[Title/Abstract] OR patient preference*[Title/Abstract] OR patient's preference*[Title/Abstract] OR patients' preference*[Title/Abstract] OR patient perspective*[Title/Abstract] OR patient's perspective*[Title/Abstract] OR patients' perspective*[Title/Abstract] OR patient's experience*[Title/Abstract] OR patients' experience*[Title/Abstract] OR "health status"[Title/Abstract] OR "health state"[Title/Abstract] OR "emotional strain"[Title/Abstract] OR "disability"[title] OR disabl*[Title] OR "unmet need*" [Title/Abstract] OR caregiver*[Title/Abstract] OR "care giver*" [Title/Abstract] OR "anxiety"[Title/Abstract] OR "fatigue"[Title/Abstract] OR "depression"[Title/Abstract])

Search number	Search terms
Clinical burden	
#4	#1 AND ("Fibromatosis, Aggressive/mortality"[Majr] OR "Fibromatosis, Aggressive/complications"[Majr] OR "Disease Progression"[Majr] OR "Comorbidity"[Majr] OR complicat*[Title] OR sequelae[Title] OR "disease progression"[Title] OR mortalit*[Title] OR death*[Title] OR fatal*[Title] OR comorbidit*[Title/Abstract] OR "co-morbidit*[Title/Abstract] OR "multi-morbidit*[Title/Abstract] OR "disease exacerbation"[Title/Abstract] OR "progression of disease"[Title/Abstract] OR "misdiagnosis"[Title/Abstract])
Economic burden	
#5	#1 AND ("Fibromatosis, Aggressive/economics"[Mesh] OR "Unemployment"[Mesh] OR "Health Care Costs"[Mesh] OR "Absenteeism"[Mesh] OR "Presenteeism"[Mesh] OR "Costs and Cost Analysis"[Mesh] OR "Economics"[Mesh] OR "Economics, Hospital"[Mesh] OR "Economics, Medical"[Mesh] OR "Economics, Nursing"[Mesh] OR "Economics, Pharmaceutical"[Mesh] OR "Budgets"[Mesh] OR "Efficiency"[Mesh] OR "Insurance, Disability"[Mesh] OR "Health Expenditures"[Mesh] OR "Cost of Illness"[Mesh] OR "Cost-Benefit Analysis"[Mesh] OR "Direct Service Costs"[Mesh] OR "Length of Stay"[Mesh] OR "Health Resources/economics"[Mesh] OR "Fees and Charges"[Mesh] OR "Quality-Adjusted Life Years"[Mesh] OR cost[Title/Abstract] OR costs[Title/Abstract] OR costly[Title/Abstract] OR economic*[Title/Abstract] OR pharmaco-economic*[Title/Abstract] OR fiscal[Title/Abstract] OR fee[Title/Abstract] OR fees[Title/Abstract] OR expenditure*[Title/Abstract] OR budget*[Title/Abstract] OR "resource use"[Title/Abstract] OR "resource utilization"[Title/Abstract] OR "resource utilisation"[Title/Abstract] OR "length of stay"[Title/Abstract] OR "quality adjusted life year"[Title/Abstract] OR "quality adjusted life years"[Title/Abstract] OR QALY*[Title/Abstract] OR absenteeism[Title/Abstract] OR productiv*[Title/Abstract] OR "lost work"[Title/Abstract] OR "work loss"[Title/Abstract] OR "missed work"[Title/Abstract] OR "sick day*[Title/Abstract] OR "illness day*[Title/Abstract] OR presenteeism[Title/Abstract] OR "q-twist"[Title/Abstract])
Treatments	
#6	#1 AND ("Fibromatosis, Aggressive/drug therapy"[Majr] OR "Fibromatosis, Aggressive/therapy"[Majr] OR "Therapeutics"[Majr] OR "Drug Therapy"[Majr] OR "active surveillance"[Title/Abstract] OR "cryotherapy"[Title/Abstract] OR "radiation"[Title/Abstract] OR "refractory"[Title/Abstract] OR "sorafenib"[Title/Abstract] OR "pazopanib"[Title/Abstract] OR "imatinib"[Title/Abstract]) AND (systematic[SB] OR review*[Title] OR "systematic review*[Title/Abstract] OR SLR[Title/Abstract] OR "literature review*[Title/Abstract] OR "Review"[Publication Type] OR "Systematic Review"[Publication Type])
Practice patterns and guidelines	
#7	#1 AND ("Practice Patterns, Physicians"[Mesh] OR "Practice Guidelines as Topic"[Mesh] OR "Practice Guideline"[Publication Type] OR treatment pattern*[Title/Abstract] OR practice pattern*[Title/Abstract] OR current treatment*[Title/Abstract] OR "treatment options"[Title/Abstract] OR best practice*[Title/Abstract] OR guideline*[Title/Abstract])
Randomized controlled trials	
#8	#1 AND ("Randomized Controlled Trial"[Publication Type] OR "Controlled Clinical Trial"[Publication Type] OR randomized[Title/Abstract] OR placebo[Title/Abstract] OR "Clinical Trials as Topic"[Mesh:NoExp] OR randomly[Title/Abstract] OR trial[Title])
Observational studies	
#9	#1 AND ("Cross-Sectional Studies"[Mesh] OR "Cohort Studies"[Mesh] OR "Case-Control studies"[Mesh] OR "Observational Study" [Publication Type] OR "Retrospective Studies"[Mesh] OR "Prospective Studies"[Mesh] OR "Registries"[Mesh] OR registr*[Title/abstract] OR "chart review"[Title/abstract] OR "administrative data"[Title/abstract] OR claims data*[Title/abstract] OR electronic medical record*[Title/abstract] OR electronic health record*[Title/abstract] OR "Electronic Health Records"[Mesh] OR "real world"[TIAB] OR "Medicare claims"[Title/abstract] OR "SEER"[Title/abstract] OR "Surveillance, Epidemiology, and End Results"[Title/abstract] OR "Surveillance, Epidemiology and End Results"[Title/abstract] OR "retrospective"[Title/abstract] OR "prospective"[Title/abstract])

Search number	Search terms
Exclusions	
#10	"Animals"[Mesh] NOT "Humans"[Mesh]
#11	"Comment"[Publication Type] OR "Letter"[Publication Type] OR "Editorial"[Publication Type] OR "Case Reports"[Publication Type]
Total	
#12	(#2 OR #3 OR #4 OR #5 OR #6 OR #7 OR #8 OR #9) NOT (#10 OR #11)

Table S2. Embase Literature Search Strategy: Burden of Illness of DT (Search Conducted 18 November 2021))

Limits: last 10 years; English only; humans; no comments, letters, editorials, or case reports

Search number	Search terms
Disease	
#1	('fibromatosis'/exp/mj OR (fibromatosis:ti AND aggressive:ti) OR 'aggressive fibromatosis':ti OR 'desmoid type fibromatosis':ti OR (fibromatosis:ti AND desmoid:ti) OR 'desmoid tumor':ti OR 'desmoid tumours':ti OR 'desmoid tumors':ti OR 'desmoid tumour':ti OR 'desmoid fibromatosis':ti OR 'deep musculoaponeurotic fibromatosis':ti OR 'musculoaponeurotic fibromatosis':ti) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Epidemiology	
#2	#1 AND ('fibromatosis'/exp/mj/dm_ep OR 'epidemiology'/exp/mj OR 'morbidity'/exp/mj OR 'incidence'/exp/mj OR 'prevalence'/exp/mj OR 'risk factor'/exp/mj OR epidemiolog*:ti OR incidence*:ti OR prevalence:ti OR morbidit*:ti OR risk*:ti) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Humanistic burden	
#3	#1 AND ('quality of life'/exp OR 'self report'/exp OR 'daily life activity'/exp OR 'patient satisfaction'/exp OR 'patient preference'/exp OR 'quality adjusted life year'/exp OR 'health status'/exp OR 'mental stress'/exp OR burden:ti,ab OR 'quality of life':ti,ab OR 'life quality':ti,ab OR 'qol':ti,ab OR 'hrql':ti,ab OR 'hrqol':ti,ab OR 'euroqol':ti,ab OR 'eq5d':ti,ab OR 'eq 5d':ti,ab OR 'quality of well being':ti,ab OR 'well being':ti,ab OR 'sf-6d':ti,ab OR 'patient report*':ti,ab OR 'proxy report*':ti,ab OR 'self report*':ti,ab OR questionnaire*:ti,ab OR 'functional status':ti,ab OR 'activities of daily living':ti,ab OR 'physical function':ti,ab OR 'patient satisfaction':ti,ab OR 'patient preference*':ti,ab OR 'patients preference*':ti,ab OR 'patient perspective*':ti,ab OR 'patients perspective*':ti,ab OR 'patient experience*':ti,ab OR 'patients experience*':ti,ab OR 'health status':ti,ab OR 'health state':ti,ab OR 'emotional strain':ti,ab OR disability:ti OR disabl*:ti OR 'unmet need*':ti,ab OR caregiver*:ti,ab OR 'care giver*':ti,ab OR 'anxiety':ti,ab OR 'fatigue':ti,ab OR 'depression':ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Clinical burden	
#4	#1 AND ('fibromatosis'/exp/mj/dm_co OR 'disease exacerbation'/exp/mj OR 'comorbidity'/exp/mj OR complicat*:ti OR sequelae:ti OR 'disease progression':ti OR mortalit*:ti OR death*:ti OR fatal*:ti OR comorbidit*:ti,ab OR 'co-morbidit*':ti,ab OR 'multi-morbidit*':ti,ab OR 'disease exacerbation':ti,ab OR 'progression of disease':ti,ab OR misdiagnosis:ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py

Search number	Search terms
Economic burden	
#5	#1 AND ('unemployment'/exp OR 'health care cost'/exp OR 'absenteeism'/exp OR 'presenteeism'/exp OR 'cost'/exp OR 'economics'/exp OR 'health economics'/exp OR 'pharmacoeconomics'/exp OR 'budget'/exp OR 'productivity'/exp OR 'disability insurance'/exp OR 'cost of illness'/exp OR 'cost benefit analysis'/exp OR 'length of stay'/exp OR 'health care utilization'/exp OR 'fee'/exp OR 'quality adjusted life year'/exp OR cost:ti,ab OR costs:ti,ab OR costly:ti,ab OR economic*:ti,ab OR pharmacoeconomic*:ti,ab OR fiscal:ti,ab OR fee:ti,ab OR fees:ti,ab OR expenditure*:ti,ab OR budget*:ti,ab OR 'resource use':ti,ab OR 'resource utilization':ti,ab OR 'resource utilisation':ti,ab OR 'length of stay':ti,ab OR 'quality adjusted life year':ti,ab OR 'quality adjusted life years':ti,ab OR qaly*:ti,ab OR absenteeism:ti,ab OR productivity*:ti,ab OR 'lost work':ti,ab OR 'work loss':ti,ab OR 'missed work':ti,ab OR 'sick day*':ti,ab OR 'illness day*':ti,ab OR presenteeism:ti,ab OR 'q-twist':ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Treatments	
#6	#1 AND ('fibromatosis'/exp/mj/dm_dt OR 'fibromatosis'/exp/mj/dm_th OR 'therapy'/exp/mj OR 'drug therapy'/exp/mj OR 'active surveillance':ti,ab OR cryotherapy:ti,ab OR radiation:ti,ab OR refractory:ti,ab OR sorafenib:ti,ab OR pazopanib:ti,ab OR imatinib:ti,ab) AND (review*:ti OR 'systematic review*':ti,ab OR slr:ti,ab OR 'literature review*':ti,ab OR 'review'/exp OR 'systematic review'/exp) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Practice patterns and guidelines	
#7	#1 AND ('clinical practice'/exp OR 'practice guideline'/exp OR 'treatment pattern*':ti,ab OR 'practice pattern*':ti,ab OR 'current treatment*':ti,ab OR 'treatment options':ti,ab OR 'best practice*':ti,ab OR guideline*:ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Randomized controlled trials	
#8	#1 AND ('randomized controlled trial'/exp OR 'controlled clinical trial'/exp OR randomized:ti,ab OR placebo:ti,ab OR 'clinical trial (topic)'/de OR randomly:ti,ab OR trial:ti) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Observational studies	
#9	#1 AND ('cross-sectional study'/exp OR 'cohort analysis'/exp OR 'case control study'/exp OR 'observational study'/exp OR 'retrospective study'/exp OR 'prospective study'/exp OR 'register'/exp OR registr*:ti,ab OR 'chart review':ti,ab OR 'administrative data':ti,ab OR 'claims data*':ti,ab OR 'electronic medical record*':ti,ab OR 'electronic health record*':ti,ab OR 'electronic health record'/exp OR 'real world':ti,ab OR 'medicare claims':ti,ab OR 'seer':ti,ab OR 'surveillance, epidemiology, and end results':ti,ab OR 'surveillance, epidemiology and end results':ti,ab OR 'retrospective':ti,ab OR 'prospective':ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Exclusions	
#10	'animal'/exp NOT 'human'/exp AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
#11	(comment*:ti OR 'letter':it OR 'editorial':it OR 'case report'/exp OR [conference abstract]/lim OR [conference paper]/lim OR [conference review]/lim OR 'conference abstract':it OR 'conference paper':it) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Total	
#12	(#2 OR #3 OR #4 OR #5 OR #6 OR #7 OR #8 OR #9) NOT (#10 OR #11)

**Table S3. Cochrane Literature Search Strategy: Burden of Illness of DT
(Search Conducted 18 November 2021)**

Limits: last 10 years; English only; humans; no comments, letters, editorials, or case reports

Search number	Search terms
Disease	
#1	[mh "fibromatosis, aggressive"[mj]] OR ((fibromatosis AND aggressive) OR "aggressive fibromatosis" OR "desmoid type fibromatosis" OR (fibromatosis AND desmoid) OR "desmoid tumor" OR "desmoid tumours" OR "desmoid tumors" OR "desmoid tumour" OR "desmoid fibromatosis" OR "deep musculoaponeurotic fibromatosis" OR "musculoaponeurotic fibromatosis"):ti
Epidemiology	
#2	#1 AND ([mh "Fibromatosis, Aggressive"[mj]/EP) OR [mh "Epidemiology"[mj]] OR [mh "Morbidity"[mj]] OR [mh "Incidence"[mj]] OR [mh "Prevalence"[mj]] OR [mh "Risk Factors"[mj]] OR (epidemiolog* OR incidence* OR prevalence OR morbidit* OR risk*):ti
Humanistic burden	
#3	#1 AND ([mh "Quality of Life" OR [mh "Self Report" OR [mh "Activities of Daily Living" OR [mh "Patient Satisfaction" OR [mh "Patient Preference" OR [mh "Quality-Adjusted Life Years" OR [mh "Health Status" OR [mh "Stress, Psychological" OR (disability OR disabl*):ti OR (burden OR "quality of life" OR "life quality" OR "QoL" OR "hrql" OR "hrqol" OR "EuroQoL" OR "EQ5D" OR "EQ 5D" OR "quality of well being" OR "well being" OR "SF-6D" OR "patient NEXT report*" OR "proxy NEXT report*" OR "self NEXT report*" OR questionnaire* OR "functional status" OR "activities of daily living" OR "physical function" OR "patient satisfaction" OR "patient NEXT preference*" OR "patient's NEXT preference*" OR "patients' NEXT preference*" OR "patient NEXT perspective*" OR "patient's NEXT perspective*" OR "patients' NEXT perspective*" OR "patient NEXT experience*" OR "patient's NEXT experience*" OR "patients' NEXT experience*" OR "health status" OR "health state" OR "emotional strain" OR "unmet NEXT need*" OR caregiver* OR "care NEXT giver*" OR anxiety OR fatigue OR depression):ti,ab,kw)
Clinical burden	
#4	#1 AND ([mh "Fibromatosis, Aggressive"[mj]/MO) OR [mh "Fibromatosis, Aggressive"[mj]/CO) OR [mh "Disease Progression"[mj]] OR [mh "Comorbidity"[mj]] OR (complicat* OR sequelae OR "disease progression" OR mortalit* OR death* OR fatal*):ti OR (comorbidit* OR "co NEXT morbidit*" OR "multi NEXT morbidit*" OR "disease exacerbation" OR "progression of disease" OR misdiagnosis):ti,ab,kw)
Economic burden	
#5	#1 AND ([mh "Fibromatosis, Aggressive"/EC) OR [mh "Unemployment" OR [mh "Health Care Costs" OR [mh "Absenteeism" OR [mh "Presenteeism" OR [mh "Costs and Cost Analysis" OR [mh "Economics" OR [mh "Economics, Hospital" OR [mh "Economics, Medical" OR [mh "Economics, Nursing" OR [mh "Economics, Pharmaceutical" OR [mh "Budgets" OR [mh "Efficiency" OR [mh "Insurance, Disability" OR [mh "Health Expenditures" OR [mh "Cost of Illness" OR [mh "Cost-Benefit Analysis" OR [mh "Direct Service Costs" OR [mh "Length of Stay" OR [mh "Health Resources"/EC) OR [mh "Fees and Charges" OR [mh "Quality-Adjusted Life Years" OR (cost OR costs OR costly OR economic* OR pharmacoeconomic* OR fiscal OR fee OR fees OR expenditure* OR budget* OR "resource use" OR "resource utilization" OR "resource utilisation" OR "length of stay" OR "quality adjusted life year" OR "quality adjusted life years" OR QALY* OR absenteeism OR productiv* OR "lost work" OR "work loss" OR "missed work" OR "sick NEXT day*" OR "illness NEXT day*" OR presenteeism OR "q-twist"):ti,ab,kw)
Treatments	
#6	#1 AND ((([mh "Fibromatosis, Aggressive"[mj]/DT) OR [mh "Fibromatosis, Aggressive"[mj]/TH) OR [mh "Therapeutics"[mj]] OR [mh "Drug Therapy"[mj]] OR ("active surveillance" OR cryotherapy OR radiation OR refractory OR sorafenib OR pazopanib OR imatinib):ti,ab,kw) AND (review*:ti OR ("systematic NEXT review*" OR SLR OR "literature review*"):ti,ab,kw OR ("Review" OR "Systematic Review"):pt))

Search number	Search terms
Practice patterns and guidelines	
#7	#1 AND ([mh "Practice Patterns, Physicians"] OR [mh "Practice Guidelines as Topic"] OR ("Practice Guideline":pt) OR ("treatment NEXT pattern*" OR "practice NEXT pattern*" OR "current NEXT treatment*" OR "treatment options" OR "best NEXT practice*" OR guideline*):ti,ab,kw)
Randomized controlled trials	
#8	#1 AND ([mh ^"Clinical Trials as Topic"] OR ("Randomized Controlled Trial" OR "Controlled Clinical Trial"):pt OR (randomized OR placebo OR randomly):ti,ab,kw OR trial:ti)
Observational studies	
#9	#1 AND ([mh "Cross-Sectional Studies"] OR [mh "Cohort Studies"] OR [mh "Case-Control studies"] OR [mh "Retrospective Studies"] OR [mh "Prospective Studies"] OR [mh "Registries"] OR [mh "Electronic Health Records"] OR (registr* OR "chart review" OR "administrative data" OR "claims NEXT data*" OR "electronic NEXT medical NEXT record*" OR "electronic NEXT health NEXT record*" OR "real world" OR "Medicare claims" OR SEER OR "Surveillance, Epidemiology, and End Results" OR "Surveillance, Epidemiology and End Results" OR retrospective OR prospective):ti,ab,kw OR ("Observational Study":pt))
Exclusions	
#10	[mh "Animals"] NOT [mh "Humans"]
#11	("Comment" OR "Letter" OR "Editorial" OR "Case Reports"):pt OR "conference abstract"
Total	
#12	(#2 OR #3 OR #4 OR #5 OR #6 OR #7 OR #8 OR #9) NOT (#10 OR #11)
#13	#12 AND Publication date from 2011/11/18

Table S4. Embase Conference Abstracts Search: Burden of Illness of DT

Limits: 2015-present; English only; humans; no comments, letters, editorials, or case reports

Search number	Search terms
Disease	
#1	('fibromatosis'/exp/mj OR (fibromatosis:ti AND aggressive:ti) OR 'aggressive fibromatosis':ti OR 'desmoid type fibromatosis':ti OR (fibromatosis:ti AND desmoid:ti) OR 'desmoid tumor':ti OR 'desmoid tumours':ti OR 'desmoid tumors':ti OR 'desmoid tumour':ti OR 'desmoid fibromatosis':ti OR 'deep musculoaponeurotic fibromatosis':ti OR 'musculoaponeurotic fibromatosis':ti) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Epidemiology	
#2	#1 AND ('fibromatosis'/exp/mj/dm_ep OR 'epidemiology'/exp/mj OR 'morbidity'/exp/mj OR 'incidence'/exp/mj OR 'prevalence'/exp/mj OR 'risk factor'/exp/mj OR epidemiolog*:ti OR incidence*:ti OR prevalence:ti OR morbidit*:ti OR risk*:ti) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py

Search number	Search terms
Humanistic burden	
#3	#1 AND ('quality of life'/exp OR 'self report'/exp OR 'daily life activity'/exp OR 'patient satisfaction'/exp OR 'patient preference'/exp OR 'quality adjusted life year'/exp OR 'health status'/exp OR 'mental stress'/exp OR burden:ti,ab OR 'quality of life':ti,ab OR 'life quality':ti,ab OR 'qol':ti,ab OR 'hrql':ti,ab OR 'hrqol':ti,ab OR 'euroqol':ti,ab OR 'eq5d':ti,ab OR 'eq 5d':ti,ab OR 'quality of well being':ti,ab OR 'well being':ti,ab OR 'sf-6d':ti,ab OR 'patient report*':ti,ab OR 'proxy report*':ti,ab OR 'self report*':ti,ab OR questionnaire*:ti,ab OR 'functional status':ti,ab OR 'activities of daily living':ti,ab OR 'physical function':ti,ab OR 'patient satisfaction':ti,ab OR 'patient preference*':ti,ab OR 'patients preference*':ti,ab OR 'patient perspective*':ti,ab OR 'patients perspective*':ti,ab OR 'patient experience*':ti,ab OR 'patients experience*':ti,ab OR 'health status':ti,ab OR 'health state':ti,ab OR 'emotional strain':ti,ab OR disability:ti OR disabl*:ti OR 'unmet need*':ti,ab OR caregiver*:ti,ab OR 'care giver*':ti,ab OR 'anxiety':ti,ab OR 'fatigue':ti,ab OR 'depression':ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Clinical burden	
#4	#1 AND ('fibromatosis'/exp/mj/dm_co OR 'disease exacerbation'/exp/mj OR 'comorbidity'/exp/mj OR complicat*:ti OR sequelae:ti OR 'disease progression':ti OR mortalit*:ti OR death*:ti OR fatal*:ti OR comorbidit*:ti,ab OR 'co-morbidit*':ti,ab OR 'multi-morbidit*':ti,ab OR 'disease exacerbation':ti,ab OR 'progression of disease':ti,ab OR misdiagnosis:ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Economic burden	
#5	#1 AND ('unemployment'/exp OR 'health care cost'/exp OR 'absenteeism'/exp OR 'presenteeism'/exp OR 'cost'/exp OR 'economics'/exp OR 'health economics'/exp OR 'pharmacoeconomics'/exp OR 'budget'/exp OR 'productivity'/exp OR 'disability insurance'/exp OR 'cost of illness'/exp OR 'cost benefit analysis'/exp OR 'length of stay'/exp OR 'health care utilization'/exp OR 'fee'/exp OR 'quality adjusted life year'/exp OR cost:ti,ab OR costs:ti,ab OR costly:ti,ab OR economic*:ti,ab OR pharmacoeconomic*:ti,ab OR fiscal:ti,ab OR fee:ti,ab OR fees:ti,ab OR expenditure*:ti,ab OR budget*:ti,ab OR 'resource use':ti,ab OR 'resource utilization':ti,ab OR 'resource utilisation':ti,ab OR 'length of stay':ti,ab OR 'quality adjusted life year':ti,ab OR 'quality adjusted life years':ti,ab OR qaly*:ti,ab OR absenteeism:ti,ab OR productiv*:ti,ab OR 'lost work':ti,ab OR 'work loss':ti,ab OR 'missed work':ti,ab OR 'sick day*':ti,ab OR 'illness day*':ti,ab OR presenteeism:ti,ab OR 'q-twist':ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Treatments	
#6	#1 AND ('fibromatosis'/exp/mj/dm_dt OR 'fibromatosis'/exp/mj/dm_th OR 'therapy'/exp/mj OR 'drug therapy'/exp/mj OR 'active surveillance':ti,ab OR cryotherapy:ti,ab OR radiation:ti,ab OR refractory:ti,ab OR sorafenib:ti,ab OR pazopanib:ti,ab OR imatinib:ti,ab) AND (review*:ti OR 'systematic review*':ti,ab OR slr:ti,ab OR 'literature review*':ti,ab OR 'review'/exp OR 'systematic review'/exp) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Practice patterns and guidelines	
#7	#1 AND ('clinical practice'/exp OR 'practice guideline'/exp OR 'treatment pattern*':ti,ab OR 'practice pattern*':ti,ab OR 'current treatment*':ti,ab OR 'treatment options':ti,ab OR 'best practice*':ti,ab OR guideline*:ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Randomized controlled trials	
#8	#1 AND ('randomized controlled trial'/exp OR 'controlled clinical trial'/exp OR randomized:ti,ab OR placebo:ti,ab OR 'clinical trial (topic)'/de OR randomly:ti,ab OR trial:ti) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py

Search number	Search terms
Observational studies	
#9	#1 AND ('cross-sectional study'/exp OR 'cohort analysis'/exp OR 'case control study'/exp OR 'observational study'/exp OR 'retrospective study'/exp OR 'prospective study'/exp OR 'register'/exp OR registr*:ti,ab OR 'chart review':ti,ab OR 'administrative data':ti,ab OR 'claims data*':ti,ab OR 'electronic medical record*':ti,ab OR 'electronic health record*':ti,ab OR 'electronic health record'/exp OR 'real world':ti,ab OR 'medicare claims':ti,ab OR 'seer':ti,ab OR 'surveillance, epidemiology, and end results':ti,ab OR 'surveillance, epidemiology and end results':ti,ab OR 'retrospective':ti,ab OR 'prospective':ti,ab) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Exclusions	
#10	'animal'/exp NOT 'human'/exp AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
#11	(comment*:ti OR 'letter':it OR 'editorial':it OR 'case report'/exp) AND [english]/lim AND [embase]/lim AND [18-11-2011]/sd AND [2011-2021]/py
Total	
#12	(#2 OR #3 OR #4 OR #5 OR #6 OR #7 OR #8 OR #9) NOT (#10 OR #11)
#13	#12 AND ([conference abstract]/lim OR [conference paper]/lim OR [conference review]/lim OR 'conference abstract':it OR 'conference paper':it) AND [2015-2021]/py
#14	#13 AND ('2015 annual meeting of the american society of clinical oncology, asco':nc OR '2016 annual meeting of the american society of clinical oncology, asco 2016':nc OR '2017 annual meeting of the american society of clinical oncology, asco':nc OR '2018 annual meeting of the american society of clinical oncology, asco 2018':nc OR '2019 annual meeting of the american society of clinical oncology, asco 2019':nc OR '42nd esmo congress, esmo 2017':nc OR '43rd congress of european society for medical oncology, esmo 2018':nc OR '44th congress of european society for medical oncology, esmo 2019':nc OR '44th esmo congress':nc OR 'annual meeting of the american society of clinical oncology, asco 2021':nc OR 'esmo congress 2021':nc OR 'esmo virtual congress 2020':nc OR 'esmo asia congress 2016':nc)

Inclusion and Exclusion Criteria

Table S5 present the inclusion exclusion criteria for reviewing studies included in this review.

Table S5. Inclusion and Exclusion Criteria: Disease Description, Epidemiology, Clinical, Economic and Humanistic Burden, Treatments and Treatment Patterns, and Guidelines

	Inclusion Criteria	Exclusion Criteria
Population	<ul style="list-style-type: none"> ▪ Patients with desmoid tumors or aggressive fibromatosis 	<ul style="list-style-type: none"> ▪ Patients with other tumors
Intervention and comparators	<ul style="list-style-type: none"> ▪ Wait and see, active surveillance, ▪ Cryotherapy, radiation ▪ Chemotherapy ▪ TKIs (Sorafenib, pazopanib, imatinib) ▪ Surgery 	<ul style="list-style-type: none"> ▪ Natural remedies
Outcomes	<ul style="list-style-type: none"> ▪ Clinical description <ul style="list-style-type: none"> – Classification – Diagnosis and misdiagnosis – Natural history – Mechanism of disease ▪ Epidemiology <ul style="list-style-type: none"> – Incidence – Prevalence – Trends – Risk factors ▪ Clinical burden <ul style="list-style-type: none"> – Mortality – Recurrence of disease progression ▪ Humanistic burden <ul style="list-style-type: none"> – Disability – Health-related quality of life – Functional status ▪ Economic burden <ul style="list-style-type: none"> – Direct costs – Indirect costs – Resource utilization ▪ Treatment patterns <ul style="list-style-type: none"> – Type of treatment used (studies evaluating surgery are of special interest) – Duration of treatment – Outcomes/efficacy – Side effects or toxicities – Discontinuations – Recurrences or disease progression 	<ul style="list-style-type: none"> ▪ Biochemical outcomes ▪ Immunological outcomes ▪ Genetic outcomes ▪ Validation

	Inclusion Criteria	Exclusion Criteria
Study design	<ul style="list-style-type: none"> ▪ Epidemiologic studies ▪ Retrospective database studies ▪ Prospective observational studies or registries ▪ Cross-sectional studies ▪ Case-control studies ▪ Cohort studies (retrospective and prospective) ▪ Practice guidelines ▪ Consensus statements ▪ Clinical trials of treatments (phase 2 or 3, focusing on TKIs) ▪ Systematic reviews/meta-analyses ▪ Focus of study: <ul style="list-style-type: none"> – Epidemiology – Clinical burden – Economic burden – Humanistic burden – Treatments or Treatment patterns – Surgery – Guidelines 	<ul style="list-style-type: none"> ▪ Small studies (n< 20) ▪ Clinical trials phase 1 ▪ Pilot studies ▪ Genetic, biomarker, diagnostic, or prognostic studies ▪ Case reports ▪ Case series (unless n >20) ▪ Editorials, letters, commentaries, expert opinion ▪ Study design, protocol only or methods paper ▪ Guideline that has been superseded by one more recent. ▪ Old reviews that have been superseded by current ones ▪ Validation studies of PROs or of biomarkers

TKI=tyrosine kinase inhibitor; PROs= patient-reported outcomes

Note: The inclusion and exclusion criteria above were applied for level 1 and level 2 screening.