Cancer Association of South Africa (CANSA)

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Fact Sheet on Desmoplastic Small-Round-Cell Tumour

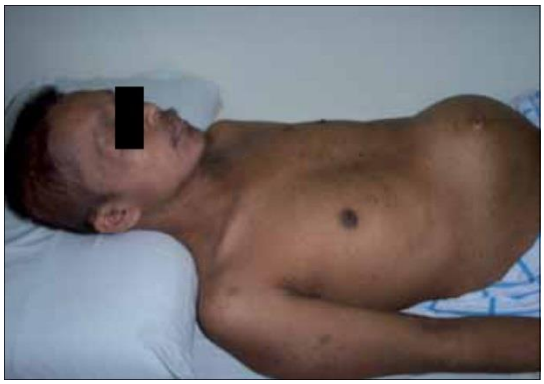
Introduction

Desmoplastic small round cell tumour (DSRCT) is generally associated with aggressive features and a poor prognosis. The prognosis for patients with DSRCT remains poor. Despite aggressive therapy, 3-year overall survival has been estimated at 44% and the 5-year survival rate remains around 15%.

DSRCT exhibits a male predominance of 90%, and 85% of patients are Caucasian. Median age at diagnosis has been reported as 14, 19 and 25 years of age in different series. The prognosis for patients with DSRCT remains poor. Despite aggressive therapy, 3-year overall survival has been estimated at 44% and the 5-year survival rate remains around 15%.

Desmoplastic Small-Round-Cell Tumour

Desmoplastic Small-Round-Cell Tumour (DSRCT) is considered a childhood cancer that predominantly strikes boys and young adults. The disease rarely occurs in females, but when it does the tumours can be mistaken for ovarian cancer. It is classified as a soft tissue sarcoma. It is an aggressive and rare tumour that primarily occurs as masses in the abdomen.



[Picture Credit: Intra-abdominal Desmoplastic Small-Round-Cell Tumour]

Other areas affected may include the lymph nodes, the lining of the abdomen, diaphragm, spleen, liver, chest wall, skull, spinal cord, large intestine, small intestine, bladder, brain, lungs, testicles, ovaries, and the pelvis. Reported sites of metastatic spread include the liver, lungs, lymph nodes, brain and bones.

Researched and Authored by Prof Michael C Herbst
[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dipt Audiometry and Sound Measurement; Diagnostic Radiographer; Medical Ethicist]
Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

Tsoukalas, N., Kiakou, M., Nakos, G., Tolia, M., Galanopoulos, M., Tsapakidis, K., Kamposioras, K., Christofyllakis, C., Dimitrakopoulos, G. & Sambaziotis, D. 2020.

"Desmoplastic small round-cell tumour is a very rare neoplasm, which usually arises from the abdominal or pelvic peritoneum of adolescents and young adults. Early diagnosis is difficult, because most tumours present with non-specific gastrointestinal symptoms after a long asymptomatic period. It is generally a very aggressive tumour, which grows rapidly with poor prognosis and an overall five-year survival rate of 15% despite multimodal treatment. Despite multiple treatment strategies, the management of desmoplastic small round-cell tumour still remains a clinical challenge and no consensus about a therapeutic protocol has been established. A 35-year-old man presented with mild abdominal pain, constipation and weight gain, and was eventually diagnosed with desmoplastic small round-cell tumour, which was shown to be limited to the abdomen. After incomplete debulking surgery, radiotherapy and chemotherapy, he developed multiple metastatic nodular foci in chest and the pleura and, unfortunately, he died due to disease progression."

Honoré, C., Delhorme, J.B., Nassif, E., Faron, M., Ferron, G., Bompas, E., Glehen, O., Italiano, A., Bertucci, F., Orbach, D., Pocard, M., Quenet, F., Blay, J.Y., Carrere, S., Chevreau, C., Mir, O., Le Cesne, A., French Network for Rare Peritoneal Malignancies (RENAPE), & French Sarcoma Clinical Network (NETSARC). 2019.

BACKGROUND: Despite being associated with a very poor prognosis, long-term survivors across all series of Desmoplastic Small Round Cell Tumor (DSRCT) have been reported.

AIM OF THE STUDY: To analyze patients 'characteristics associated with a prolonged survival after DSRCT diagnosis.

METHODS: All consecutive patients treated for DSRCT in nine French expert centers between 1991 and 2018 were retrospectively analyzed. Patients with a follow-up of less than 2 years were excluded and cure defined as being disease-free at least 5 years.

RESULTS: 100 pts were identified (median age 25 years, 89% male). 27 had distant metastases at diagnosis and 80 pts underwent upfront chemotherapy (CT). 71 pts were operated, 20 pts without prior CT). Surgery was macroscopically complete (CC0/1) in 50 pts. Hyperthermic intraperitoneal Chemotherapy (HIPEC) was administered during surgery in 15 pts 54 pts had postoperative CT and 26 pts had postoperative whole abdomino-pelvic RT (WAP-RT). After a median follow-up of 103 months (range 23-311), the median overall survival (OS) was 25 months. The 1- year, 3-year and 5-year OS rates were 90%, 35% and 4% respectively. 5 patients were considered cured after a median disease-free interval of 100 months (range 22-139). Predictive factors of cure were female sex (HR = 0.49, p = 0.014), median PCI<12 (HR = 0.32, p = 0.0004), MD Anderson stage I (HR = 0.25, p < 0.0001), CC0/1 (HR = 0.34, p < 0.0001), and WAP-RT (HR = 0.36, p = 0.00013). HIPEC did not statistically improve survival.

CONCLUSION: Cure in DSRCT is possible in 5% of patients and is best achieved combining systemic chemotherapy, complete cytoreductive surgery and WAP-RT. Despite aggressive treatment, recurrence is common and targeted therapies are urgently needed.

Incidence of Desmoplastic Small-Round-Cell Tumour

The National Cancer Registry (2016) does not provide any statistics regarding Desmoplastic Small-round-Cell Tumour in South Africa.

Risk Factors and Causes of Desmoplastic Small Round Cell Tumour (DSRCT)

There aren't any known risk factors associated with Desmoplastic Small Round Cell Tumours. The disease is known to pop up from primitive cells during childhood. Chromosomal translocations chromosome 11 and chromosome 22 causes this disease. When this happens, the body is no longer able to suppress tumour growth.

Scheer, M., Vokuhl, C., Blank, B., Hallmen, E., von Kalle, T., Münter, M., Wessalowski, R., Hartwig, M., Sparber-Sauer, M., Schlegel, P.G., Kramm, C.M., Kontny, U., Spriewald, B., Kegel, T., Bauer, S., Kazanowska, B., Niggli, F., Ladenstein, R., Ljungman, G., Jahnukainen, K., Fuchs, J., Bielack, S.S., Klingebiel, T., Koscielniak, E. & Cooperative Weichteilsarkom Studiengruppe [CWS]. 2019.

BACKGROUND: To evaluate optimal therapy and potential risk factors.

METHODS: Data of DSRCT patients <40 years treated in prospective CWS trials 1997-2015 were analyzed.

RESULTS: Median age of 60 patients was 14.5 years. Male:female ratio was 4:1. Tumors were abdominal/retroperitoneal in 56/60 (93%). 6/60 (10%) presented with a localized mass, 16/60 (27%) regionally disseminated nodes, and 38/60 (63%) with extraperitoneal metastases. At diagnosis, 23/60 (38%) patients had effusions, 4/60 (7%) a thrombosis, and 37/54 (69%) elevated CRP. 40/60 (67%) patients underwent tumor resection, 21/60 (35%) macroscopically complete. 37/60 (62%) received chemotherapy according to CEVAIE (ifosfamide, vincristine, actinomycin D, carboplatin, epirubicin, etoposide), 15/60 (25%) VAIA (ifosfamide, vincristine, adriamycin, actinomycin D) and, 5/60 (8%) P6 (cyclophosphamide, doxorubicin, vincristine, ifosfamide, etoposide). Nine received high-dose chemotherapy, 6 received regional hyperthermia, and 20 received radiotherapy. Among 25 patients achieving complete remission, 18 (72%) received metronomic therapies. Three-year event-free (EFS) and overall survival (OS) were 11% (±8 confidence interval [CI] 95%) and 30% (±12 CI 95%), respectively, for all patients and 26.7% (±18.0 CI 95%) and 56.9% (±20.4 CI 95%) for 25 patients achieving remission. Extra-abdominal site, localized disease, no effusion or ascites only, absence of thrombosis, normal CRP, complete tumor resection, and chemotherapy with VAIA correlated with EFS in univariate analysis. In multivariate analysis, significant factors were no thrombosis and chemotherapy with VAIA. In patients achieving complete remission, metronomic therapy with cyclophosphamide/vinblastine correlated with prolonged time to relapse.

CONCLUSION: Pleural effusions, venous thrombosis, and CRP elevation were identified as potential risk factors. The VAIA scheme showed best outcome. Maintenance therapy should be investigated further.

Signs and Symptoms of Desmoplastic Small-Round-Cell Tumour (DSRCT)

Desmoplastic small-round-cell tumour (DSRCT) is a rare disease of children, adolescents and young adults, which begins in the abdominal cavity. Because of the rarity of this disease, little is known about optimal treatment.

These aggressive cancers often form as multiple tumours in the tissue (peritoneum) that lines the inside of the abdomen and pelvis. They quickly spread to other structures within the abdomen.

Patients may present with dozens to hundreds of tumours studding the peritoneal cavity (abdominal cavity). Despite this presentation, it is not primarily considered metastatic but multifocal. It can metastasise to the liver or lung.

Chemotherapy, radiotherapy, and surgical approaches have not been standardised. Neoadjuvant chemotherapy often yields a partial response; however, tumours may remain surgically un-resectable.

An aggressive approach to treatment is required to maximise long-term remission.

Symptoms of desmoplastic small round cell tumour include:

- Pain or a lump in the abdomen
- Cramping
- Nausea
- Vomiting
- Diarrhoea
- Constipation
- Trouble having a bowel movement and/or passing gas
- Abdominal swelling
- Back pain
- Gastrointestinal blockage
- Lack of appetite
- Weight loss
- Fatigue
- Fluid in the abdomen (ascites)
- Anaemia
- Thyroid or hormone problems

Diagnosis of Desmoplastic Small Round Cell Tumour (DSRCT)

Diagnostic tests vary, based on location of the tumour. It is often noticed that at diagnosis, the tumour has metastasized to distant organs. Diagnosis of Desmoplastic Small Round Cell Tumour is made using the following tools:

- Physical examination, evaluation of patient's medical history
- Histopathological studies conducted on a biopsy specimen - the specimen is examined under a microscope by a pathologist, to arrive at a definitive diagnosis
- MRI, CT, and PET scans of the affected regions - to aid in obtaining a clear image of the tumour prior to surgery. A solid and firm mass is usually noticed

Many clinical conditions may have similar signs and symptoms. Your healthcare provider may perform additional tests to rule out other clinical conditions to arrive at a definitive diagnosis.

Saltsman, J.A. 3rd, Price, A.P., Goldman, D.A., Hammond, W.J., Danzer, E., Magnan, H., Slotkin, E., Tap, W.D., Heaton, T.E., Modak, S. & LaQuaglia, M.P. 2020.

Background: Desmoplastic small round cell tumor (DSRCT) is an aggressive soft tissue sarcoma affecting children and young adults with 5-year overall survival (OS) of approximately 20%. Despite generally poor prognosis, long-term survival does occur. However, no evidence-based system exists to risk-stratify patients at diagnosis.

Methods: We retrospectively reviewed all DSRCT cases diagnosed at our institution between January 2000 and September 2016. Demographics, diagnostic imaging, and clinical data were reviewed. Univariate and multivariate Cox proportional hazard modeling was used to evaluate associations between imaging characteristics and OS.

Results: There were 130 patients (85% male; median age at presentation: 21.2 years) with confirmed DSRCT and sufficient imaging and clinical information for analysis. Median 5-year OS was 28% (95% CI: 19%-37%). In univariate analysis, shorter OS was associated with presence of liver lesions (hazard ratio [HR] 2.1, 95% CI: 1.28-3.45), chest lesions (HR 1.86, 95% CI: 1.11-3.1), and ascites (HR 1.69, 95% CI: 1.06-2.7). In multivariate analysis, liver involvement and ascites were predictive and were used to stratify risk (intermediate=no liver involvement or ascites; high=either liver involvement or ascites;

very high=both liver involvement and ascites). Intermediate-risk patients had a 5-year survival of 61% (95% CI: 40%-76%) versus 16% (95% CI: 6%-29%) among high-risk patients and 8% (95% CI: 1%-29%) among very high risk patients.
Conclusion: Patients with DSRCT can be risk-stratified at diagnosis based on specific imaging characteristics.
Type of study: Retrospective study with no comparison group.
Level of evidence: Level IV.

Differential diagnosis of Desmoplastic Small-Round-Cell Tumour (DSRCT)
The following conditions are kept in mind when making a diagnosis:

- peritoneal carcinomatosis
- non Hodgkin lymphoma
- malignant peritoneal mesothelioma
- rhabdomyosarcoma
- round cell sarcomas
- Ewing sarcoma/primitive neuroectodermal tumour
    - alveolar rhabdomyosarcoma
    - desmoplastic round cell tumour
    - mesenchymal chondrosarcoma
    - poorly differentiated synovial sarcoma
- lymphomas
- neuroendocrine carcinoma
- neuroblastoma and variants

(Lessnick, *et al*, 2009)

Treatment of Desmoplastic Small-Round-Cell Tumour (DSRCT)
Because DSRCT is so rare, no standard way to treat it has been developed. The following treatment methods have been used:

- Surgery - is used to remove as much of the cancer as possible. Often, DSRCTs have spread too far for complete removal, but surgeons try to remove at least 90 percent of them
- Hyperthermic intraperitoneal chemotherapy (HIPEC) - may be given during the surgery to kill cancer cells that cannot be removed surgically. (The patient also avoids the side effects of standard chemotherapy.)
    - HIPEC is done by circulating a heated, sterile chemo solution through the part of the abdomen where the tumours are found, for up to two hours
- Chemotherapy - uses powerful medicines to kill cancer cells or stop them from growing (dividing) and making more cancer cells
    - Chemotherapy may be injected into the bloodstream, so that it can travel throughout the body
    - Some chemotherapy may be given by mouth
    - Some chemotherapy may be given by mouth

- Radiation therapy - uses high-energy X-rays or other types of radiation to kill cancer cells or stop them from growing
    - External radiation uses machines outside the body to deliver the X-ray dose.
    - Internal radiation uses needles, seeds, wires or catheters to deliver the radiation directly into or close to the cancer.

DSRCT located outside the abdomen without any spread seems to respond better to treatment than DSRCT in the abdomen or than DSRCT which has spread into other parts of the body.

DSRCT is a very aggressive neoplasm with a 5-year survival of less than 15%.

Treatment options include surgery, radiotherapy, chemotherapy with or without stem cell transplantation, and recently introduced molecularly targeted therapies. Unfortunately there is no standard therapeutic regimen described since no modality is clearly superior to any other. Surgery is usually extensive and often includes excision of the omentum, splenectomy and lymph node resections. Due to the invasive nature of this tumour, complete resection with negative margins is usually not possible. Debulking surgery has been described as an attempt to eliminate 90% of the tumour bulk.

In addition to surgery and radiation therapy, local control options for DSRCT (particularly metastatic disease) may include radiofrequency ablation, gammaknife, cryoablation, embolisation and chemoembolisation. These are usually performed in academic centres after careful consideration of individual cases.

Treatment Options

Tumour Location	Treatment Options
Peritoneal Disease	Surgery, whole abdominal radiotherapy, continuous hyperthermic peritoneal perfusion
Liver Metastases	Surgery, stereotactic radiosurgery, radiofrequency ablation, cryoablation, 90Y-microspheres
Lung Metastases	Surgery, stereotactic radiosurgery
Mediastinal Lymph Nodes	Radiation therapy
Bone Metastases	Radiation therapy

High-dose chemotherapy with autologous stem-cell rescue has also been attempted, however no significant impact in long-term survival has been achieved after transplant.

Although DSRCTs are generally sensitive to chemotherapy, the response is not enough to achieve cure since patients almost invariably relapse. This could potentially be a reflection of the heterogeneity of the cells within the tumour; where a distinct population of cells ("cancer stem cells") that are less sensitive to chemotherapy and radiotherapy possess the ability to self-renew and retain the capacity to regenerate the tumour bulk after it has been eradicated. This represents a highly attractive hypothesis since it could explain tumour behaviour and lead to the identification of new targets for more effective therapies. Unlike other small round blue cell tumours like Ewing's sarcoma, such a stem cell has not been yet identified in DSRCTs.

Stiles, Z.E., Murphy, A.J., Anghelescu, D.L., Brown, C.L., Davidoff, A.M., Dickson, P.V., Glazer, E.S., Bishop, M.W., Furman, W.L., Pappo, A.S., Lucas, J.T. Jr. & Deneve, J.L. 2020.

Background: Desmoplastic small round cell tumor (DSRCT) is a rare intra-abdominal soft tissue sarcoma affecting adolescents and young adults. Cytoreduction, hyperthermic intraperitoneal chemotherapy (CRS/HIPEC), and adjuvant radiotherapy may improve local control. We review our experience with patients who undergo CRS/HIPEC and adjuvant radiotherapy for DSRCT.
Methods: A retrospective review was performed for patients with DSRCT from 2013 to 2017 who underwent CRS/HIPEC. Clinicopathologic, operative, and outcome data were reviewed.
Results: Ten CRS/HIPEC procedures were performed for nine patients (7 males, 6 Caucasian, median age 19 years (range 10-24)). Four patients presented with extra-abdominal disease; five had liver involvement. The median peritoneal cancer index was 16 (range 5-20). All received neoadjuvant chemotherapy. CCR 0/1 resection was possible in nine patients. Major complications occurred in four with no operative mortalities. All received adjuvant chemotherapy, seven received radiation therapy, and three received stem-cell transplant. All but one patient recurred after treatment. The median recurrence-free and overall survival (OS) were 12 and 45 months (95% confidence interval 35.1-54.9) respectively, with a 3-year OS of 55%. Long-term parenteral nutrition was required in eight for a median of 261 days (range 37-997). Clinically significant long-term complications requiring further surgery included gastroparesis (N = 1), small bowel obstruction (N = 3) and hemorrhagic cystitis (N = 2).
Conclusions: Multimodal therapy for DSRCT consisting of multiagent neoadjuvant chemotherapy, CRS/HIPEC, adjuvant chemotherapy, and radiation therapy is associated with potential cumulative toxicity. Recurrence after resection is common. Prolonged parenteral nutrition may be necessary, and late gastrointestinal and genitourinary complications may require additional treatment.

Ambar, N.B.D., de Seixas Alves, M.T., Lewderman, H.M., Abib, S., Duarte, A.A.B. & Caran, E.M. 2019.
INTRODUCTION: Desmoplastic small round cell tumor is an extremely rare and aggressive cancer that affects mainly adolescents and young adults. Despite multiple therapeutic strategies, most patients have resistant disease with very poor survival rates.
CASE PRESENTATION: We present a case of a 10-year-old Caucasian boy with a desmoplastic small round cell tumor refractory to conventional treatment who exhibited a good response to alternative treatment. With use of irinotecan and vincristine in association with radiation therapy, a reduction of 96.9% of the dimensions of the target lesions compared with the initial image was observed.
CONCLUSION: This chemotherapy regimen, in association with radiation therapy, demonstrated efficacy for refractory desmoplastic small round cell tumor in our patient, and it is cost-effective.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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http://sarcomahelp.org/dsrct.html

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