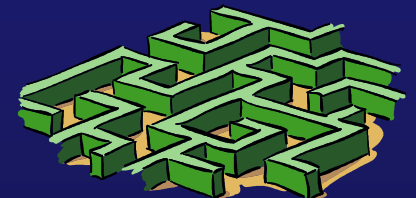


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Long term survival of Congenital
Fibrosarcoma, about 3 cases .

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Summery

- ▶ **Congenital / Infantile fibrosarcoma (C/IFS)** is a rare tumour most often affecting the extremities of **newborns, infants and young children**.
- ▶ Considering age, surgery of primary is often preferred to chemotherapy and surgical extirpation alone has therefore resulted in an excellent prognosis.
- ▶ **The amputation rate, however, is too frequent .** **The preoperative chemotherapy** can therefore avoid amputation and furthermore be useful in treatment of metastatic recurrence and his role must be emphasised .

We presented here **the management of 3 cases of CF** and review of literature



Patients and methods: patient n°1

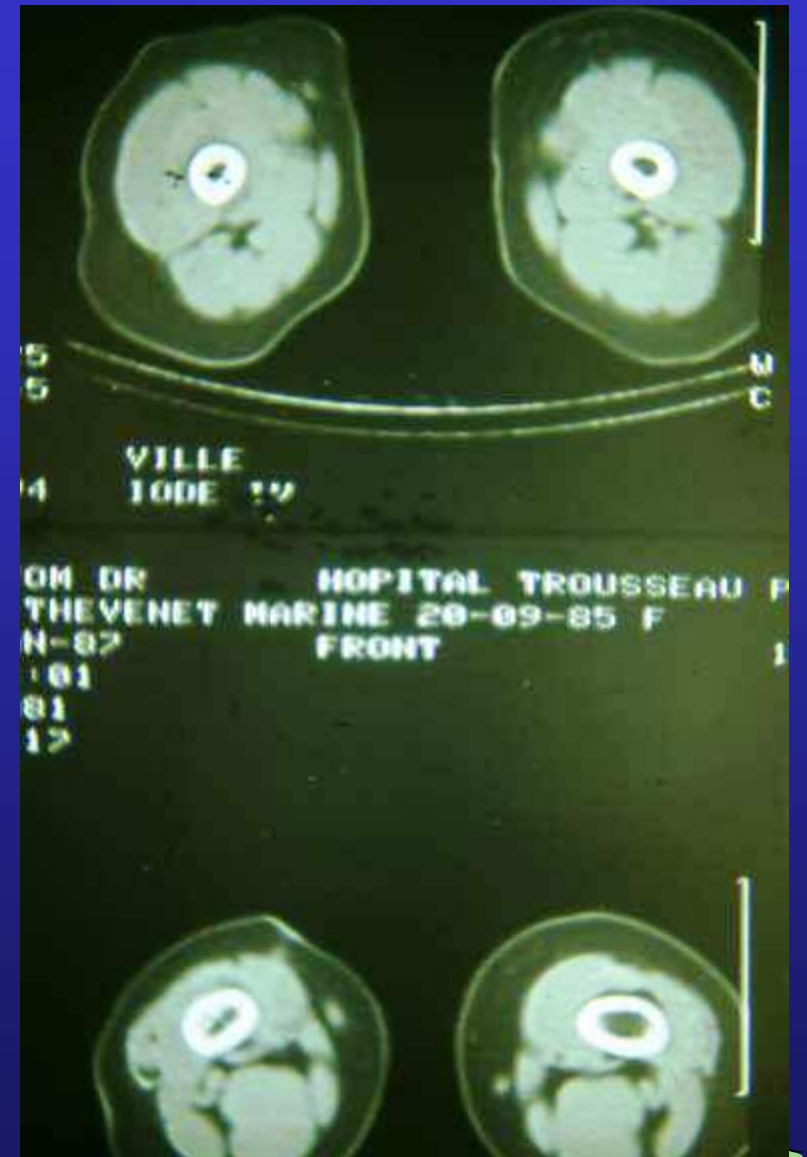
■ In 1985 , we treated a 3 months age girl for congenital fibrosarcoma of the leg.

■ To avoid amputation , we made a preoperative chemotherapy



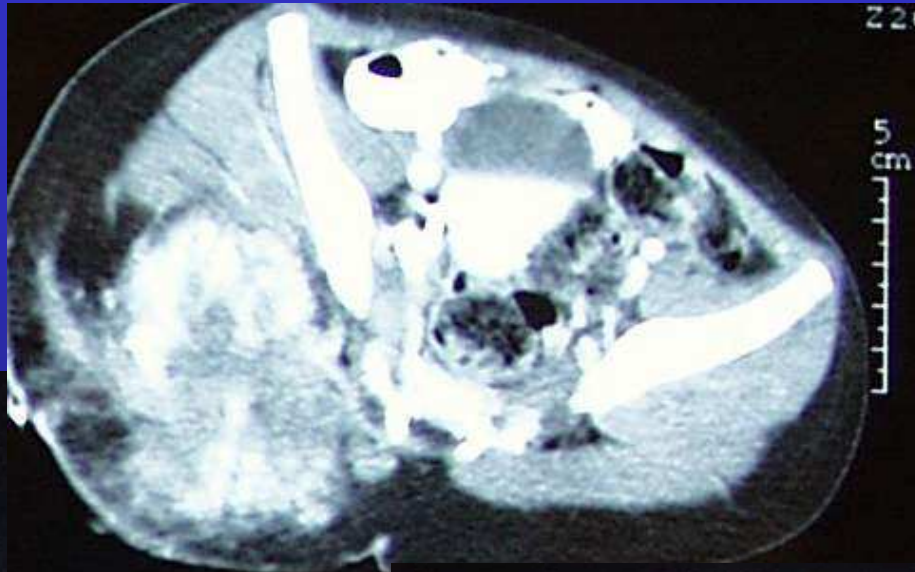
Patients and methods: patient n°1

- Chemotherapy with 3 courses of IVA at low dose in regard of the small age of the patient, with a very good local response.
- She could have a conservative surgery and we made 2 course of IVA in adjuvant treatment. She was actually in first complete remission, **25 years after the beginning of the disease.**



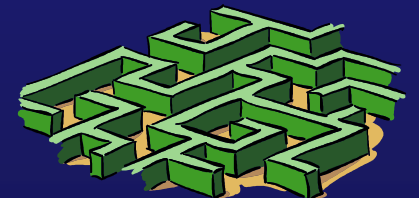
Patients and methods: patient n°2



 In september 1999, we took in care a 3 ½ years old boy for a recurrent locoregionnal buttock fibrosarcoma



Patients and methods: patient n°2

- He was previously treated elsewhere by incomplete surgery at the age of 6 months for a tumor evolving since the birth and a second time at the age of 2 years to complete the surgery.
- A new biopsy confirmed the diagnosis .
- He received 3 courses of preoperative chemotherapy by IVA with good clinical and radiological response.
- The surgery consisted in a “en-bloc extratumoral resection” with persistence of viable tumoral cells histologically






-  We completed treatment by 2 others courses of IVA and one IV (without Actinomycine D because hepatic toxicity during the last cure) .
-  In january 2003, 3 years after the end of chemotherapy, he presented bilateral pulmonary metastasis treated by surgery and postoperative chemotherapy (Cpx-Zavedos) till september 2003, time when a local recurrence appeared.

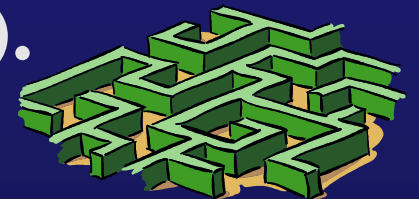


- He was submitted to a novel local surgery and post-operative chemotherapy (Cpx-Thpadriamycine, Topotecan, Doxetaxel).
- We stopped all chemotherapy in july 2004 when he developed a severe chickenpox. From this time till now , he received Alpha interferon, actually in decreased dose. He remained in third complete remission.



Patients and methods: patient n°3

-  In december 2005, we admitted **a 14 years old girl**, with a local recurrence of congenital fibrosarcoma, treated elsewhere at the age of 5 months by partial surgery and chemotherapy. She remained in first remission for 13 years.
-  Previously, in november 2005 , she received 2 courses of IPA, resulting in ulcerated tumor in the skin and the surgery could be only partial and contaminated.
-  We made two courses of chemotherapy (IVA , IV) in postoperative period , without local response and a second incomplete surgery on February 2006, followed by chemotherapy (IVA and IFX).



■ She achieved a third surgery in May 2006 , consisting on “En-Bloc complete resection”. The histologicals pathways confirmed the absence of tumor necrosis. We pursued chemotherapy by Cpx/cpx-Theprubicine/IVA/4 courses of Cpx-Hycantin, till december 2006.

■ In january 2007 , she presented a new local recurrence treated by Vindesine-Deticene , partial surgery in april 2007, post-operative radiotherapy and chemotherapy by Temodal-Campto/ taxol till august 2007.



The treatment was continued by low dose of oral CPX till january 2008, partial surgery in january 2008 , Carboplatine-Vp16 until july 2008 with partial local response, Imatinib (in the hypothesis of Dahriers Ferrand disease) for one month and tentative of 2 courses yondelis in september 2008.

Finally she undergown to an ultimate surgery consisting of amputation in november 2008.

During all theses years of local recurrence she didn't developed metastasis.



Chemotherapy for Infantile Fibrosarcoma (IF)

Review of literature

Since the 1980s **Chemotherapy** has been suggested in order to reduce the need for **extensive surgery** for IF.

In 1986, in the *Cancer* Review (1986 Oct 1;58(7):1400-6), **Ninane J** 3 cases of IF are reported. The use of preoperative chemotherapy, a **VAC regimen**, allowed conservative surgery in two of them.



Kynaston J.A presented in 1993 (*Med Pediatr. Oncol 1993;21(7):488-93*) , 9 IF who received chemotherapy : All with **Vincristine** and **Actinomycin D** , and six with additional drugs as **Ifosfamide** , **Cyclophosphamide** , **Adriamycin** , **Etoposide** , and **Cisplatinum** and six initially.

Thus, 5/9 children achieved a CR and 3/9 with chemotherapy alone. With the inclusion of chemotherapy as part of their treatment, five children, for whom curative surgery may have resulted in amputation, remain alive with limbs intact.



In the *Med Pediatr. Oncol* 1999;oct ; 33(4);
425-7, *Shetty Ak* precise also the role of
chemotherapy in the treatment of
infantile fibrosarcoma.



In the *Journal Pediatr Surg* 2000 Jun;35(6) :
Kurkchubasche AG, describe a dramatic
response in 2 recent cases to preoperative
chemotherapy, given in an attempt to avoid
amputation.

In the *Am J Périnatol*, 2008 Dec 9, **Gûlham B**,
describe the case of an unusual presentation (
pelvic mass) of IF in a male newborn who
received chemotherapy by **Vincristine** ,
Actinomycine D , **Cyclophosphamide** but but
died from veino-occlusive disease soon after
the second cycle.



Discussion and conclusion

- In our experience , like in the literature , it's seems to be indicated to treat CF with the optimum treatment especially complete en-bloc resection and preoperative chemotherapy to avoid amputation or contained resection in spite of the low age of the patients.
- This therapeutic approach can avoid local and/or metastasis recurrence like in our patient n° 1.
- Preoperative chemotherapy is feasible despite low age of the patients, with adapted dose and special care for newborns.

