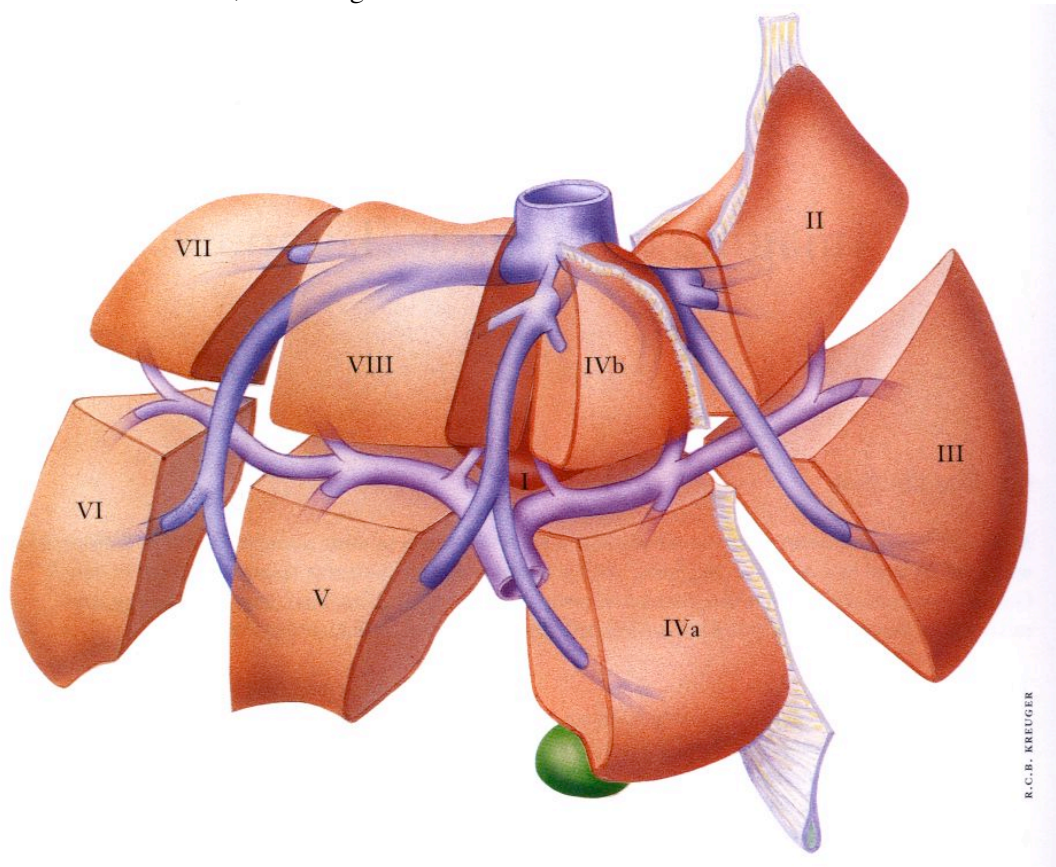


International co-operation in the SIOPEL Liver Tumours Strategy Group

Primary hepatic tumours in children are rare. They account for only 0.5 – 2% of all childhood malignancies, and for 10% of abdominal malignancies in children. 80-90% of these tumours excrete α -foetoprotein, which can be used as a means of diagnosis, and for monitoring the treatment response and recurrence.

The radical surgical removal of the tumour is the cornerstone in the curative treatment of malignant hepatic tumours in children. Accurate assessment of the lesion is crucial for the timing of surgery. Unresectable tumours may become resectable if preoperative chemotherapy is applied.

In the literature, considerable attention has been paid to surgical anatomy pertaining to liver resection. Soyer's nomenclature of segmental hepatic anatomy is accepted worldwide. He combined that of Couinaud, based on cadaver livers *ex vivo*, with those of Goldsmith & Woodburne and Bismuth. All hepatic segments, except for the caudate lobe, are defined by three vertical scissurae, and a single transverse scissura.



In 1989, Anton Vos, professor in paediatric surgery in Amsterdam, presented a pre-operative staging system in childhood hepatoblastoma, which was accepted by SIOP (International Society of Paediatric Oncology). This staging system forms the basis for the randomised clinical trials designed and carried out during the last 10 years by the SIOPEL Childhood Liver Tumour Strategy Group (SIOPEL-1 – 4, and IRINOTECAN). It is used as an assessment system of the extent of disease by radiological investigations, prior to treatment. Standard risk (SR) and high risk (HR) patients are identified. SR: those in whom the tumour has not spread outside the liver and involves, in the liver itself, either one, two, or three of the four sections of the liver. HR: those patients with tumour throughout the whole of the liver, or whose tumour has spread beyond the confines of the liver, and all HCC patients. The trials serve to evaluate different treatment protocols, and include the formulation of stopping rules, the formation of an expert panel for rapid consultation on staging and resectability, and initially the central review of the imaging material. Other objectives of these studies are:

- to further validate the pre-treatment tumour extension system (PRETEXT)
- to evaluate the feasibility of the rapid central review
- to evaluate the role of orthotopic liver transplantation for patients whose tumour remains unresectable after chemotherapy
- to evaluate an appropriate salvage therapy for relapsed patients.

The Group is backed by a Trial Office, housed in Leicester (UK), and consists of paediatric oncologists and surgeons, a paediatric radiologist, a pathologist, a statistician, and a secretary. There is an extensive e-mail contact between the members of the group. Regularly, small and larger conferences are held to discuss progress and problems of current trials, and designing new ones.

A website is maintained, containing all necessary information for participants and other doctors, and for parents of patients (<http://siopel.org>). The designing and writing of scientific articles based on the results of the trials is done in a co-operative way, depending on the predominant discipline involved, and on the character of the journal in question. The list of authors is fairly made according to the weight of each individual contribution.

Working in international groups may lead, apart from advanced patient care, to improved contact and better understanding between colleagues of different disciplines, systematic education, and to international co-operation even of doctors with an individualistic mind. Great fun!

Some publications and abstracts of (members of) the Group:

Aronson DC, Schnater JM, Staalman CR, Weverling GJ, Plaschkes J, Perilongo G, Brown J, Phillips A, Otte JB, Czauderna P, Mackinlay G, Vos A. Predictive value of the Pretreatment Extent of disease system in hepatoblastoma: Results from the International Society of Pediatric Oncology Liver Tumor Study Group SIOPEL-1 Study. *J Clin Oncol*. 23: 1245-52, 2005

Perilongo G, Shafford E, Maibach R, Aronson D, Brugieres L, Brock P, Childs M, Czauderna P, MacKinlay G, Otte JB, Pritchard J, Rondelli R, Scopinaro M, Staalman C, Plaschkes J. Risk-adapted treatment for childhood hepatoblastoma: final report of the second study of the International Society of Paediatric Oncology-SIOPEL 2. *Eur J Cancer* 40: 411-21, 2004

Otte JB, Pritchard J, Aronson DC, Brown J, Czauderna P, Maibach R, Perilongo G, Shafford E, Plaschkes J. Liver transplantation for hepatoblastoma: results from the International Society of Pediatric Oncology (SIOP) study SIOPEL-1 and review of the world experience. *Pediatr Blood Cancer* 42: 74-83, 2004

P. Czauderna, G. Mackinlay, G. Perilongo, J. Brown, E. Shafford, D. Aronson, J. Pritchard, P. Chapchap, J. Keeling, J. Plaschkes, J.B. Otte. Hepatocellular Carcinoma in Children: Results of the First Prospective Study of the International Society of Pediatric Oncology Group. *J Clin Oncol* 20:2798-2804, 2002

Schnater JM, Aronson DC, Plaschkes J, Perilongo G, Brown J, Otte JB, Brugieres L, Czauderna P, MacKinlay G, Vos A. Surgical view of the treatment of patients with hepatoblastoma: results from the first prospective trial of the International Society of Pediatric Oncology Liver Tumor Study Group. *Cancer* 94: 1111-20, 2002

Aronson DC, Schnater JM, Plaschkes J, Perilongo G, Brown J, Staalman CR, Czauderna P, Otte JB, Mackinlay G, Vos A. Surgical aspects of the treatment of hepatoblastoma - Experience of the first prospective study (SIOPEL-1) (ab). *Med Pediatr Oncol* 37: 193, 2001

Plaschkes J, Kellie S, Czauderna P, Maibach R, Shafford E, Staalman C, Childs M, Brugières L, MacKinlay G, Perilongo G. SIOP Childhood Tumour Study Group: SIOPEL - Annual overview report on hepatoblastoma Clinical research (ab). *Med Pediatr Oncol* 37: 239, 2001

A copy of the 2005 article, including the PRETEXT system, is handed out during the meeting.