Our Experience with the Haemophiliac in Need for Surgery

Results in Connection with Bleeding Haemophiliacs in the Course of 1 Year of Activity

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The necessity for surgical interventions has for many years been considered as a serious risk for the haemophiliac. Small surgery, however (such as sutures of superficial wounds, incision of abscesses, etc.) could be done rather easily but the surgery of internal organs was performed only when an emergency forced the issue.

Such difficulties have been prevalent in our country until not so long ago. Laparotomy for emergency abdominal conditions was considered by the surgeon with a great deal of skepticism, even when large quantities of anti-haemophilic plasma and/ or frozen plasma were available. The increasing number of emergencies in haemophiliacs determined the search for a solution, and this is reflected in the present study.

The problems related to the laboratory diagnosis were solved by the production of special reagents, as well as by the introduction of new devices, and organizational aspects have been dealt with by preparing complete files for a total of 607 patients with type A, and for 73 with type B haemophilia.

The most important problem was that of cryoconcentrate production.

This was started at a modest rate in 1972, when only 80 flaks were prepared, but the production attained nearly 2,000 flaks in 1979. Production of cryoconcentrate was also started in some other blood collection and transfusion centres in the country.

The introduction of the cryoconcentrate made possible the application of surgical procedures for haemophiliacs, and this is done in a unified manner in the whole country, where the same schemes for the therapy have been recommended in all the clinics where haemophiliacs are hospitalized.

During 1979 we have been requested to provide assistance in 138 medical and 22 surgical problems regarding haemophiliacs.

The medical cases included haemarthroses with various localizations (predominantly of the knees), as well as haematuria, epistaxis, spontaneous haematomas, meningeal haemorrhages and cerebral compression, and cerebral haematoma. The surgical cases consisted of fractures, post-traumatic wounds, appendicitis, hernia, peritonitis, stomatological and orthopaedic interventions. We have started by carefully assessing the patients, using multiple tests also aimed

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at detecting possible organic deficiencies besides the coagulation defect. Thus we had to introduce additional therapeutic measures in some of the patients for hepatic affections (that were more numerous), but also for cardiopathies and digestive diseases. In the moderately severe

cases (haemarthroses, epistaxis, haematuria), the solution was easy to find, while in other cases we encountered some difficulties, mostly in relation to the severity of the symptoms, and to the development, in the polytransfused patients, of antibodies against the coagulation factors, so that the amounts that had to be administered had to be considerably increased.

Another problem we were confronted with was that of the latent fibrinolysis, rather frequently encountered in our patients. This was solved by giving EACA in doses of 300 mg/kg of body weight administered in three daily doses under laboratory control, before, during and after surgery.

Special problems were raised by: – 1 case of meningeal haemorrhage associated with cerebral compression; – a perirenal haematoma suggesting an abdominal emergency, and haematomas located in the anterior abdominal wall suggesting also an abdominal emergency.

As a rule, evaluation of the haematomas was avoided because the infection risk had to be considered, as well as the negative effects of rapid decompression. Another unsolved problem is that of the development of a biologically unexplained haemorrhage during some orthopaedic interventions (synovectomy), when the procedure was repeated for the 4th or 5th time. These are probably due to local fibrinolysis phenomena.

The general scheme for treatment was

based on the following formula:

concentration of factor VIII or of factor IX desired to be achieved $\times$ body weight (in kg):

The number of units administered are equally divided over 24 h at 8-hour intervals.

The efficiency of the treatment was controlled by testing the prothrombin consumption time, by the cephalin-kaolin time, and more rarely by a dosage of the factor implicated. The treatment was started 8 h before surgery, and in emergencies within 1–2 h before surgery. Treatment was prolonged in relation to the evolution and healing process. The duration of the treatment was as short as possible (in contrast with some other authors), because we have attempted to prevent the development of anti-factor antibodies.

With regard to type B haemophilia we did not have any particular problems with those patients who were treated with anti-haemophilic plasma, frozen plasma, etc. Beginning with the next year, small amounts of PPSB will probably be produced in our laboratory.

Conclusions

We are satisfied by the fact that the bleeding haemophiliac does no longer not represent a serious problem, considering the experience that was acquired by our laboratory with regard to the amounts of anti-haemophilic preparations to be administered, as well as the additional therapeutic measures that seemed necessary. Our cooperation with the various clinical units has been very satisfactory. There are still problems in some departments where the cryo-

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concentrate is not produced as yet. Anti-haemophilic plasma is available in these departments, however, and the production of an isothermal container is currently considered to transport the cryoconcentrate by air.

We consider that the fact of making possible major surgery in haemophiliac patients is our greatest achievement. This is especially true for complex orthopaedic surgical procedures, which are currently performed in two