

'Why Haemophilia?'

The last 25 years have witnessed phenomenal growth in the understanding and treatment of haemophilia and other congenital bleeding disorders. In the years following World War II, a plethora of new clotting factors was described and the formation of a clot appeared to be an incomprehensible, complex interaction of innumerable factors. In 1953 an international meeting was convened to standardize nomenclature. The result was surprisingly successful and the meeting served to simplify both the nomenclature of the clotting factors and the number of their interactions. The congenital bleeding disorders were thought, at that time, to be due to deficiencies of specific clotting factors. At the same time, blood fractionation techniques were developed to prepare plasma and plasma products in large quantities. This allowed close study of the treatment of the congenital bleeding conditions, and the development of rational therapeutic regimens.

However, the incidence of these disorders is low, and outside the larger centres, the chances of the individual patient receiving the benefits of the therapeutic advances were often not great. Through the vision and motivation of one Canadian haemophiliac, Mr Frank Schnabel, an effort was made to organize patients and interested medical personnel into an international body whose aims and objects were to provide a fellowship for sufferers from haemophilia and allied disorders, to provide a forum for reporting research and advances in treatment, and to stimulate the improvement of treatment facilities in those countries in which they were suboptimal. Mr Schnabel's dream has grown into the vigorous and active World Federation of Haemophilia (WFH), of which he is the Chairman.

This organization now comprises of haemophilia societies from 38 countries. The South African Haemophilia Foundation was elected a member in 1970. The WFH has liaison with other international organizations such as the World Health Organization, the International Society for the Rehabilita-

tion of the Disabled, and the Council of World Organizations Interested in the Handicapped.

In Mr Schnabel's chairman's report delivered at the VIIIth Congress of the WFH held in Teheran, Iran, on 17 - 20 May 1971, he asked the question 'Why haemophilia?' Mr Schnabel then stated:

'Why haemophilia when there are more pressing medical and social problems? Recently the WFH received a letter from a doctor in a developing country, who is exploring the possibility of organizing a haemophilia society in his country. However, the decision has been deferred because haemophilia is not severe enough in view of other priorities like malnutrition, sickle-cell anaemia, etc.

'Let me preface my observations by saying, of course we are deeply concerned with malnutrition, but malnutrition is such an overwhelming problem it boggles the mind. Haemophilia, in contrast, is a relatively minor health problem, but therein lies its **attraction to action**. Definite and specific progress can be achieved fairly quickly. Haemophilia lends itself to a prototype and the experience gained can be applied to other health defects. Secondly, the concurrent attack on multiple health problems can be complementary. As a concept in logic on humanism, we can hardly await the solution to malnutrition before turning to haemophilia. Thirdly, a strong case can be made that it requires more medical attention and more hospitalization costs **not** to organize and **not** to treat haemophilia with the contemporary methods available. The usual crisis-to-crisis mismanagement of haemophilia can be more expensive than a "control" management of haemophilia. Then, how do we assess the disruption in the family caused by a disabled haemophiliac, or the burden to the community of a welfare case?

'But less obvious and less well known is the constructive influence haemophilia has had on other aspects of medicine:

1. Interest in haemophilia has been paramount in the development of knowledge of blood coagulation which has, in turn, advanced thrombosis research.

2. Blood from haemophiliacs was the first and still is the richest source of antibodies to the virus of serum (transfusion) hepatitis. Thus, haemophilia has directly and crucially advanced hepatitis research.

3. Haemophilia treatment, so dependent upon blood transfusion facilities, has given great impetus to the concept of component therapy and has thus advanced the quality of blood transfusion practices.

'Haemophilia offers a unique prototype for the partnership of doctor and patient in the assault

on an illness, for the establishment of multispecialty organization for the comprehensive care of one disorder, for the study of the influence of sociologic development upon the natural history of one disease, and for consideration of the worldwide implications of a serious genetic disorder with a high mutation rate.

'There are rather few combined patient-doctor endeavours on the international scene. In our wanderings about the globe, on more than one occasion, we've been told "we are watching you, if you succeed in this joint effort then other health defects should travel the same road".'

Die Federale Raad en die Pers

Justice must not only be done; it must be seen to be done.

As mens niks het om oor skaam te wees nie, het jy ook niks om weg te steek nie. Dit geld as breë beleid, maar daar is natuurlik sekere aspekte van die doen en late van enige organisasie wat nie aan die publiek beskikbaar gestel kan word nie. Sou dit wenslik of wys wees om die vergaderings van die Federale Raad van die Mediese Vereniging van Suid-Afrika aan die pers oop te stel?

Die Geneeskundige en Tandheekkundige Raad se vergaderings moet noodwendig oop wees aangesien dit 'n statutêre liggaam is. Van tyd tot tyd is dit egter ook vir die Mediese Raad nodig om **in camera** te debatteer, en al wat in sulke gevalle nodig is, is dat 'n voorstel aanvaar word dat die Raad in komitee gaan. Dit is belangrik om op te merk dat die Mediese Raad die toelating van die publiek as so belangrik beskou dat enige mosie om in komitee te debatteer, behoorlik gemotiveer moet wees. Dieselfde geld in ons howe. Alvorens 'n hof **in camera** sitting kan hou, moet daar afdoende rede voor wees en so 'n versoek word nie sommer ligtelik toegestaan nie.

Die notule van die vergaderings van die Federale Raad word in die **Tydskrif** gepubliseer en is daar vir enigeen ter insae. Waarom stel ons dan nie ook die debat aan die publiek oop nie? Indien dit geen ander vrugte afwerp nie, sal dit ten minste tot gevolg hê dat die koerantberigte oor die mediese beroep op beter feitekennis gegrond sal wees. Wanneer ons na die toesprake in die Federale Raad luister, kan ons nie anders nie as om oortuig te wees van die opregte pogings wat daar aangewend word om die beste moontlike mediese organisasie tot stand te bring. Kom ons gee aan die publiek, ons pasiënte, die geleentheid om hulself hiervan te kom oortuig.

Die Raad sal natuurlik nou en dan, of selfs

kort-kort as dit daarop aankom, in komitee moet gaan en enige persman sal dit eenvoudig moet aanvaar, maar die grootste gedeelte van die debat kan oop wees en dit sal sonder twyfel leersaam wees vir diegene wat tog so gereed is met skietgoed teen die Vereniging. As daar miskien hier en daar 'n lid van die Federale Raad is wat soms geneig is om ondeurdagte dinge te sê, sal dit moontlik dien om hom in toom te hou as hy weet dat sy stoomafblasery miskien die volgende oggend voorbladnuus sal wees.

Die Mediese Vereniging was nog nooit 'n geheime organisasie nie en mag dit ook nooit word nie. Ons is as dokters daar om die mensdom te dien, nie om agter geslote deure of in donker hoekies te beraadslaag nie. Iedere praktiserende geneesheer gebruik daagliks sy oordeel om te besluit hoeveel hy aan 'n gegewe pasiënt moet meedeel en hoeveel liefs agterweë gehou moet word, maar geen dokter gaan lank die vertrouwe van sy pasiënte behou as hy konsekwent weier om enige inligting te verstrek nie. Daardie oordeel insake die weerhouding van sekere feite kan in die Raad uitgeoefen word deur **in camera** te debatteer.

Ons is 'n tradisievolle beroep en die eerste reaksie sal natuurlik onvermydelik 'nee!' wees; bloot omdat ons nog al die jare nie die pers of die publiek toegelaat het nie. 'n Mens moet egter waak dat tradisie nie die goeie organisasie begin oorheers nie. As ons nog nooit die Federale Raad se notules gepubliseer het nie, sou daar seker sterk reaksie gewees het as iemand so 'n voorstel sou maak het.

Ons **Tydskrif** is aan iedereen beskikbaar, ons notules word gepubliseer, kom ons stel nou ook ons vergaderings oop.