EDITORIAL : VAN DIE REDAKSIE

THROMBOASTHENIA

In some haemorrhagic states the abnormal bleeding cannot be attributed to defective coagulation or to thrombocytopenia. Many of these states appear to be primary, inborn, inherited diseases, the inheritance being that of an autosomal dominant so that both males and females are affected. Numerous cases may occur in one family. In many cases the haemorrhage is purpuric in type and the clinical syndrome does not differ very markedly from that seen in thrombocytopenic states. The platelet count is normal, but there is sometimes a long bleeding time and/or a positive reaction to the tourniquet test. On the basis of these findings it is not unreasonable that our attention should be focussed more closely both on the capillaries and on the blood platelets. The normality of the platelet count has led to the conclusion that if there is a platelet abnormality it must be one of function—a type of thromboasthenia.

The names which have been given to these haemorrhagic conditions are legion. Constitutional thrombopathy, thromboasthenia, pseudohaemophilia, and Von Willebrand-Jürgens syndrome are a few examples. In discussing these conditions it is helpful to consider them under two broad headings, though there may well be overlapping between the groups. There are those where the lesion seems to be in the capillaries, and those in which thromboasthenia appears to be present. Some patients have both types of defect. The most classical example of a capillary disorder is hereditary capillary telangiectasia, but this condition is generally regarded as a separate entity. In the capillary disorder considered here, the only laboratory abnormalities detected have been a prolonged bleeding time with or without demonstrable abnormalities in the capillaries, and possibly with positive tourniquet tests. Cases with a similar clinical pattern have, however, been described where the long bleeding time has been associated with a host of coagulation defects.[1] These deficiencies have involved antihaemophilic globulin (AHG), Christmas factor, factors V and VII, and fibrinogen as well as combinations of these.

The most interesting of these conditions (and probably the most common) is the one associated with AHG deficiency. A curious feature of cases with AHG deficiency is that the clinical picture is usually not that of haemophilia. This has given rise to the rather disturbing concept that the absence of AHG does not necessarily imply a diagnosis of haemophilia. The difference in severity of the clinical picture can possibly be explained by the fact that in the capillary disorder the AHG level has usually been recorded as more than 1 - 2 per cent, while in classical haemophilia the AHG level is frequently less than this. Haemarthrosis has only rarely been recorded in the capillary disorder and is common in haemophilia. There appears, however, to be no good reason why combined defects of this type with little or no circulating AHG should not occur, and this would give rise to the clinical syndrome of 'haemophilia with long bleeding time'. Indeed, Valberg and Brown[1] go so far as to assert that some cases reported as 'haemophilia in the female' are really of this type. This may be correct in some of the reported cases. It is probably not true in all. It is also of interest to note that the disease originally described by Von Willebrand and Jürgens in the Aaland islands is of the type associated with some deficiency of circulating AHG.[2]

The group of qualitative platelet defects is even more difficult to define. Glanzmann[3] was the first to make this suggestion, and it was stated that the agglutination of platelets was delayed by the use of a 'thrombometer'. Budtz-Olsen[4] suggested that many of Glanzmann's patients actually had true thrombocytopenia, but despite this there are still grounds for the concept of defective platelet function. Platelet functions are not easy to study because at least eight clotting and haemostatic factors have been described as occurring in platelets.[5] In 38 cases of what they called 'thrombopathia' van Creveld *et al.*[5] found a varying picture of platelet dysfunction. In general more than one platelet function was affected, and the impaired platelet functions varied from case to case. In addition, in some patients the dysfunctions varied from time to time and different combinations occurred at different times. In one large family with slight familial thrombopathia, the dysfunctions varied from patient to patient. It is clear that the diagnostic criteria laid down by Glanzmann, viz. normal bleeding time and deficient clot retraction, cannot be strictly applied to this group. Van Creveld *et al.* found patients with evidence of 'thrombopathia' with prolonged bleeding times and normal clot retraction. Recently too, additional evidence of platelet dysfunction in the Willebrand-Jürgens type of thrombopathia has been reported. A 'platelet osmotic resistance test' has been devised. The standard thromboplastin-generation test is used, but the concentration of NaCl in the diluting fluid used for platelets is varied. Some patients' platelets are reported to show resistance to disintegration even in distilled water.[6]

Both 'groups' may, therefore, be associated with a prolonged bleeding time and a positive tourniquet test; there may be evidence of a host of plasma coagulation defects in the one variety and evidence of qualitative platelet deficiency in the other.

Treatment of these cases is not very satisfactory, but fortunately they frequently present in a mild form. It is important to make the correct diagnosis if only to avoid operations such as splenectomy which are usually of little or no value. It is also important to recognize that the condition is different from true haemophilia. Surgery in true haemophilia is still a hazardous procedure; it is, however, less dangerous in cases with capillary disorders. If plasma factors are missing, then judicious replacement may be of value in correcting this plasma defect, though even this correction has not always occurred.[7] The long bleeding time is generally unaffected. Freshly prepared platelet transfusions in plastic bags may be of temporary benefit

but are not generally recommended. Perhaps we may have a more definitive treatment for this interesting group of diseases in the future.

1. Valberg, L. S. and Brown, G. M. (1958): Medicine (Baltimore), 37, 181.
2. Jürgens, R., Lehmann, W., Wegelins, O., Eriksson, A. W. and Hiepler, E. (1957): Thromb. diath. haem. (Stuttg.), 1, 257.
3. Glanzmann, E. (1918): Jb. Kinderheilk., **88**, 113.
4. Budtz-Olsen, O. E. (1951): *Clot Retraction*, p. 87. Oxford: Blackwell.
5. Van Creveld, S., Liem, K. H. and Veder, H. A. (1958): Acta haemat. (Basel), **19**, 199.
6. Ulutin, O. N. and Karaca, M. (1959): Brit. J. Haematol., **5**, 302.
7. Biggs, R. and MacFarlane, R. G. (1957): *Human Blood Coagulation and its Disorders*, p. 296. Oxford: Blackwell.

DIE GEVARE VAN NARKOSE IN DIE VERLOSKUNDE

Daar word gesê dat die toediening van narkose in die verloskunde meer gevare as enige andere operatiewe ingreep inhou. Hierdie probleem is belangrik omdat nie alleen die moeder nie, maar ook die ongebore baba aan onderdrukkende verdowingsmiddels onderwerp word — soms deur onervare persone op die gebied van die narkose.

Operatiewe kraam word dikwels onder baie moeilike omstandighede beoefen, en die grootste bedreiging vir die vrou en die baba lê juis hierin dat 'n narkose in 'n plek toegedien word waar daar geen suierstelle of soortgelyke geriewe vir die verwydering van braaksel bestaan nie. Dit is algemeen bekend dat 'n vrou in kraam, selfs na ses uur, nog nie haar kos verteer het nie, en hierdie onverteerde kos bly 'n bedreiging vir die lugweg van die pasiënt en dus ook indirek vir die baba.

Selfs al word kraam onder ideale omstandighede beoefen, is daar nog altyd die onbekende uitwerking van die verdowingsmiddels op die baba. Voor die geboorte sal die sirkulasie van die baba alles absorbeer wat aan die moeder toegedien word. In hierdie opsig moet daarop gelet word dat selfs veslappingsmiddels tot die baba se sirkulasie kan deurdring en dus sy asemhaling na geboorte kan onderdruk.[1]

Hoe kan ons dan hierdie toedrag van sake verhelp? Daar bestaan geen twyfel dat die hoë sterftesyfer wat onder moeders voorkom aan die inaseming van braaksel onder algemene narkose toegeskryf kan word nie. Die oplossing van hierdie probleem lê nie voor die hand nie. Dit is onprakties om 'n kraampasiënt te belet om te eet, of om 'n vloeistofdieet voor te skryf. Waar 'n algemene narkose toegedien moet word, is 'n wye maagbuis noodsaaklik. Die klein buisie van Ryle is absoluut nutteloos. Daar is 'n toenemende neiging om net 'n trageale buis onder narkose in te sit, maar dit is seer sekerlik nie die beste manier om te voorkom dat die maag se inhoud in die longe beland nie. 'n Beter oplossing sou wees om lokale of koudale verdowing vir hierdie onvoorbereide gevalle te gebruik. Waar toestande nie ideaal is nie, behoort nie meer as 'n lae tangverlossing gedoen te word nie, en hiervoor is die pudendale blok sekerlik die ideale narkose.

Dit is essensieel dat die dosis van verdowingsmiddels gedurende kraam en narkose ingekort word en dat die toediening van verslappingsmiddels, veral van Flaxedil, streng beperk word. Gardiner[2] het daarop gewys dat Flaxedil deurdring tot die fetus se sirkulasie, maar dat d-tubokurarien dit nie doen nie. Die wenslikheid om tiopentoon aan die kraampasiënt toe te dien, berus op die toestand van die ongebore baba asook die van die moeder. Crawford[1] wys daarop dat die toediening van 'n maksimum van 250 mg. tiopentoon nie onwenslik is nie, maar dat dit na 45 sekondes in die baba se sirkulasie voorkom en dat die konsentrasie na tien minute heelwat verminder. Daar word dus voorgestel dat as tiopentoon gebruik word daar nie gehaas moet word nie, want 'n mate van versuim sal meer perifere distribusie van die tiopentoon veroorsaak en dus help om 'n lewendige baba met 'n aktiewe respiratoriese sentrum voort te bring.

Ten laaste behoort ons ons bevolking meer en meer op te voed om van inrigtings en hospitale vir hul bevallings gebruik te maak. Ons behoort ook die staat aan te moedig om meer en meer kraambeddens in die hospitale beskikbaar te stel. As ander lande dit kan doen, waarom nie Suid-Afrika nie?

1. Crawford, J. S. (1956): Brit. J. Anaesth., **28**, 201.
2. Crawford, J. S. en Gardiner, J. E. (1956): *Ibid.*, **28**, 154.