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EDITORIAL

SURGERY IN HEREDITARY HAEMORRHAGIC STATES

Patients who have a personal or family history of an abnormal tendency to bleed are often a source of anxiety when surgery is contemplated. The patient may assert that he (or she!) is a haemophilic and may present good documentary evidence of his tendency to continue bleeding after minimal trauma. What attitude should be adopted in these instances?

It is never safe to belittle such a story. The fact that a similar story has been ignored in other patients without their coming to grief is no safeguard. On the other hand if the patient has had his haemorrhagic diathesis since birth and has undergone a major surgical procedure without severe haemorrhage he almost certainly is not a true haemophilic and it is unlikely that he will come to much harm. But, unless a careful history is taken, sooner or later a severe bleeder will be encountered on the operating table with dire consequences. Even if all laboratory tests are normal a cautious attitude should be maintained and, while surgery may be undertaken under these conditions, the surgeon should be prepared to perform an emergency blood-transfusion if it becomes necessary. The patient's blood-group must be determined beforehand and blood must be available for immediate use. Despite all modern refinements of diagnosis many bleeders still exist in whom no laboratory abnormality can be demonstrated. Indeed, they constitute the majority of bleeder patients. The patient's history of severe bleeding in the past should be credited despite the absence of laboratory confirmation. Fortunately there is not much difficulty in the diagnosis of the two important conditions—haemophilia and christmas disease.

In these two conditions surgery is especially hazardous even when the patient is one of the so-called 'mild haemophilics'.[1] In this hazardous group, too, one must include patients with circulating anticoagulants,[2] many of whom will be haemophilics. Let no one delude himself that he will always be able to stop bleeding by mechanical means. Many surgeons believe that adequate compression will always ensure haemostasis. This is not true in haemophilia. The real

VAN DIE REDAKSIE

SNYKUNDE BY ERFLIKE HEMORRAGIESE TOESTANDE

Pasiënte met 'n persoonlike of familiegeskiedenis van 'n abnormale neiging om te bloei, is dikwels 'n bron van kommer wanneer 'n operasie oorweeg word. Die pasiënt mag beweer dat hy (of sy!) 'n bloeier is en mag goeie dokumentêre bewyse voorlê van sy neiging om ná 'n geringe besering aan te hou met bloei. Watter houding behoort in hierdie gevalle aangeneem te word?

Dit is nooit veilig om so 'n storie te verkleineer nie. Die feit dat 'n soortgelyke storie by ander pasiënte verontagsaam is sonder dat hulle iets oorgekom het is geen waarborg nie. Aan die anderkant, as die pasiënt sy hemorragiese diatese van geboorte af gehad het en 'n groot operasie sonder ernstige bloeding ondergaan het, is dit amper seker dat hy nie 'n egte bloeier is nie en dit is onwaarskynlik dat hy iets sal oorkom. Maar tensy 'n sorgvuldige geskiedenis verkry word, sal daar vroeër of later 'n ernstige bloeier op die operasietafel teengekom word en die gevolge sal ontsettend wees. Al is ál die laboratoriumtoetse normaal, moet sorg deurgaans uitgeoefen word en, terwyl snykunde onder hierdie toestande onderneem mag word, behoort die snydokter gereed te wees om, indien nodig, 'n noodbloedoortapping uit te voer. Die pasiënt se bloedgroep moet vooraf bepaal word en bloed moet vir onmiddellike gebruik beskikbaar wees. Afgesien van alle moderne verfynde metodes van diagnose, is daar nog baie bloeiers in wie geen laboratorium-abnormaliteit gedemonstreer kan word nie. Inderdaad hulle vorm die meerderheid van bloeierpasiënte. Die pasiënt se geskiedenis van hewige bloeding in die verlede moet geglo word ten spyte van die afwesigheid van bevestiging deur die laboratorium. Gelukkig is dit nie baie moeilik om die twee belangrike kondisies—hemofilie en christmassiekte—te diagnoseer nie.

Snykunde by hierdie twee kondisies is veral gewaagd, al is die pasiënt een van die sogenaamde *'mild haemophilics'*.[1] By hierdie gevaarlike groep moet ook pasiënte met sirkulerende stollingsteenmiddels[2] ingesluit word. Baie van hierdie pasiënte sal bloeiers wees. Laat niemand hom self mislei nie dat hy altyd in staat sal wees om die bloeding deur meganiese middels te keer nie. Baie snydokters glo dat genoegsame druk altyd bloedstelping sal verseker. Dit geld nie in hemofilie nie. Die werklike gevaar vir 'n bloeiende hemofilie-lyer lê daarin dat spanning agter die meganiese versperring sal opbou. Druknekrose ná 'n gesnyde palm mag tot totale weefselsterfte lei, ná besnyding tot gangreen van

danger in the haemophilic who bleeds is that tension will build up behind the mechanical barrier. Pressure necrosis after a cut palm may lead to gross death of tissue; after circumcision, to gangrene of the penis, and so on. A typical example is furnished by extraction of teeth. If the socket is tightly plugged or the edges of the gum are sewn together, the bleeding may go on and the blood seep into the soft tissue around the neck and the patient may bleed to death into his tissues or suffocate. The correct treatment[3] is to plug the socket lightly (using local haemostatics if one is so inclined), apply a previously prepared dental splint, and treat the patient's haemorrhagic state by means of blood or plasma transfusion, if possible before, during and after the operation. Fresh blood or plasma is required in classical haemophilia. Stored blood or serum is preferable in christmas disease. The quantity of blood needed to fully correct the coagulation defect is very large—two or three pints may be as a drop in the ocean. Preparations of anti-haemophilic globulin (AHG) at present available in this country are for the most part inferior to fresh blood. A potent preparation prepared from bovine and porcine plasma has been produced by the Oxford workers,[4] but it is not yet generally available. It has already proved its worth.[5] Unfortunately it tends to be antigenic and has caused severe anaphylactic reactions as well as thrombocytopenia; and this may severely limit its use. For these reasons elective surgery in haemophilia must be kept to an absolute minimum, especially where the abdomen or chest needs to be opened. If any one has any doubts on the severity of bleeding which may continue despite massive transfusion let him read some of the published accounts.[6,7] Surgery is a little safer when the bleeding, if it continues, will be external. There are published accounts of gastrectomy in haemophilia,[8] and even splenectomy has been done, but there are almost certainly many unpublished cases of death after such surgery.

Operation notes in haemophilia often follow a common pattern. The surgeon is surprised at the paucity of bleeding on the operation table. Haemostasis is easily secured. Vessels are tied and do not bleed. The operating field is dry and the patient leaves the table in good condition. An hour or two later bleeding starts and is continuous. The reason for this is that there is nothing wrong with the capacity of the vessels to contract after injury. This secures haemostasis. Once the spasm relaxes, and no adequate clot has formed to take its place, bleeding starts again.

The surgeon may comfort himself with the view that most abdominal emergencies in haemophilia are caused by haemorrhage into the wall of the bowel; that the so-called acute appendix in haemophilia is most times a subserous haematoma of the caecum and will subside with conservative therapy; and even should it be a true case of acute appendicitis the mortality today with antibiotics and blood and plasma transfusion is probdie peester, ensovoorts. Die trek van tande verskaf 'n tipiese voorbeeld. As die tandholte styf toegestop word, of die kante van die tandvleis toegenaai word, mag die bloeding aanhou en in die sagte weefsel rondom die nek syfer, en die pasiënt mag in die weefsels doodbloei of verstik. Die regte behandeling[3] is om die holte liggies toe te stop (terwyl plaaslike bloedstelpmiddels aangewend word, as 'n mens daartoe geneë is), gebruik te maak van 'n vooraf bereide tandsplint, en om, indien moontlik, die pasiënt se hemorragiese toestand by wyse van bloed- of plasma-oortapping voor, gedurende of ná die operasie te behandel. Vars bloed of plasma is vir klassieke hemofilie nodig. Opgegaarde bloed of serum is verkieslik vir christmas-siekte. Die hoeveelheid bloed nodig om die stollingsgebrek volkome te bowe te kom, is baie groot—twee of drie pinte mag soveel soos 'n druppel in die oseaan wees. Preparate van anti-hemofiliese globulien (AHG) tans in ons land verkrygbaar, is meesal van minder waarde as vars bloed. 'n Sterk preparaat wat van bees- of varkplasma voorberei word, is deur Oxford-navorsers[4] vervaardig, maar dit is nog nie algemeen beskikbaar nie. Dit het alreeds sy waarde bewys.[5] Ongelukkig is dit geneig om antigenies te wees, en het dit hewige anafilaktiese reaksies sowel as trombositopenie veroorsaak; dit mag die gebruik daarvan baie beperk. Om hierdie redes moet hemofilie-gevalle wat vir snykunde gekeur word tot 'n absolute minimum beperk word, veral waar die buik of bors oopgemaak moet word. Indien enigeen die minste twyfel het oor die hewigheid van bloeding wat, tenspyte van grootskaalse oortapping, voortduur, behoort hy sommige van die gepubliseerde verslae[6,7] te lees. Snykunde is ietwat veiliger as die bloeding, indien dit aanhou, uitwendig is. Daar is gepubliseerde verslae van gastrektomie wat op hemofiliese gevalle uitgevoer en selfs splenektomie wat gedoen is, maar dit is byna seker dat baie verslae van sterfgevalle ná sulke operasies nie gepubliseer is nie.

Operasienotas by hemofilie volg dikwels 'n algemene patroon. Die chirurg is verbaas oor die geringheid van bloeding op die operasietafel. Bloedstelping word maklik verseker. Bloedvate word gebind en bloei nie. Die operasie-oppervlakte is droog en die pasiënt verlaat die tafel in 'n goeie toestand. Bloeding begin na 'n uur of twee en is aanhoudend. Die rede hiervoor is dat daar niks verkeerd is met die vermoë van die vate om na besering saam te trek nie. Dit verseker bloedstelping. Na verslapping van die sametrekking begin bloeding weer as 'n doeltreffende klont nie gevorm was nie om die plek van die sametrekking in te neem nie.

Die snydokter mag homself troos met die mening dat by hemofilie die meeste buiknoodgevalle deur bloeding in die dermwand veroorsaak word; dat die sogenaamde akute blindederm by hemofilie meestal 'n subsereuse hematoom van die blindederm is en dat dit met konserwatiewe behandeling sal sak, en al sou dit 'n egte geval van blindedermontsteking wees, is die sterftesyfer vandag, met behulp van antibiotika, bloed- en plasma-oortapping, waarskynlik nie hoër ás 10% nie, as dit so hoog is. Die sterftesyfer vir buiksnykunde by hemofiliese gevalle is waarskynlik hoër as 50%. Tensy die chirurg tevrede is dat die sterftesyfer sonder

ably not greater than 10%, if it is as much as that. Mortality of abdominal surgery in haemophilia probably exceeds 50%.[6] Unless the surgeon is satisfied that the mortality without surgery is greater than that he will be advised to let well alone.

The position is far less grim when one turns to consider the other groups of hereditary bleeding diathesis. In most of these surgery is fairly safe. As a corollary to this it is usually right to say that one may correctly operate in a case of hereditary bleeding diathesis presenting in a female. Fresh or stored blood, plasma or serum, may help to correct the bleeding state and allow the surgery to be performed. Vitamin K or Vitamin K1 may be needed if there is 'hypoprothrombinaemia', though these are only rarely of value. Fibrinogen is used for the odd case of fibrinogenopenia. It can be stated categorically that calcium plays no part in the treatment of any hereditary haemorrhagic diathesis described to date. In the absence of overt scurvy it is extremely doubtful if vitamin C is of any value. Flavinoids and adrenochromes have been disappointing. Many trade preparations exist whose names appear to indicate their ability to exert a favourable influence on blood coagulation. Their exotic names represent practically their only claim to efficacy for, almost without exception, they are worthless. Provided haemophilia and related conditions have been excluded by the sensitive thromboplastin-generation test it is usually safe to go ahead with essential (contrasted with cosmetic) surgery. The definition of 'essential' might perhaps be a little more stringent than that usually adopted.

As with other diseases, close cooperation between the pathologist, physician and surgeon is essential if the patient is to receive the best treatment at present available.

1. Merskey, C. (1951): Brit. Med. J., **1**, 906.
2. Editorial (1956): S. Afr. Med. J., **30**, 723 (4 August).
3. Macfarlane, R. G. and Biggs, R. (1955): *The Diagnosis and Treatment of Haemophilia and Related Conditions*, p. 21. London: H.M. Stationery Office.
4. Macfarlane, R. G., Biggs, R. and Bidwell, E (1954): Lancet, **1**, 1316.
5. Fraenkel, G. J. and Honey, G. E. (1955): *Ibid.*, **2**, 1117.
6. Craddock, C. G., Fenninger, L. D. and Simmons, B. (1948): Ann. Surg., **128**, 888.
7. Schiller, F., Neligan, G. and Budtz-Olsen, O. E. (1948): Lancet, **2**, 842.
8. Walker, W. (1955): *Ibid.*, **1**, 749.

snykunde hoër as dit is, is dit raadsaam om nie te opereer nie.

Die posisie is nie so onverbiddelik nie as die ander groepe erflike bloeiende diatese betrag word. Snykunde is betreklik veilig in die grootste gedeelte van hierdie groepe. Hiervan kan afgelei word dat dit gewoonlik korrek is om te sê dat 'n mens met reg kan opereer in 'n geval van erflike bloeiende diatese wanneer die pasiënt 'n vrou is. Vars of opgegaarde bloed, plasma of serum mag help om die bloeding te bemeester sodat die operasie uitgevoer kan word. Vitamine-K of Vitamine-K1 mag nodig wees as daar ‚hipoprotrombinemie' is, alhoewel hulle maar selde van waarde is. Fibrinogeen word vir die enkele geval van fibrinogenopenie gebruik. Dit kan kategories gestel word dat kalsium geen deel het nie in die behandeling van enige erflike hemorragiese diatese wat tot op datum beskryf is. As skeurbuik nie definitief aanwesig is nie, is dit hoogs twyfelagtig of Vitamine-C van enige waarde is. Flavinoïede en andrenochrome was teleurstellend gewees. Daar bestaan baie handelspreparate die name waarvan skynbaar hulle vermoë aantoon om 'n gunstige invloed op bloedstolling uit te oefen. Hulle eksotiese name verteenwoordig feitlik hulle enigste aanspraak op doeltreffenheid, omdat hulle amper sonder uitsondering, waardeloos is. Mits hemofilie en verwante kondisies deur die sensitiewe tromboplastien-generasietoets uitgesluit is, is dit gewoonlik veilig om met essensieële (in teenstelling met kosmetiese) snykunde voort te gaan. Die definisie van ‚essensieële' behoort miskien ietwat strenger te wees as wat gewoonlik aanvaar word.

Soos met ander siektes, is noue samewerking tussen die patoloog, internis en snydokter noodsaaklik sodat die pasiënt die beste behandeling wat vandag beskikbaar is, kan ontvang.

1. Merskey, C. (1951): Brit. Med. J., **1**, 906.
2. Van die Redaksie (1956): S. Afr. T. Geneesk., **30**, 723 (4 Augustus).
3. Macfarlane, R. G. en Biggs, R. (1955): *The Diagnosis and Treatment of Haemophilia and Related Conditions*, bl. 21. Londen: H.M. Stationery Office.
4. Macfarlane, R. G., Biggs, R. en Bidwell, E. (1954): Lancet, **1**, 1316.
5. Fraenkel, G. J. en Honey, G. E. (1955): *Ibid.*, **2**, 1117.
6. Craddock, C. G., Fenninger, L. D. en Simmons, B. (1948): Ann. Surg., **128**, 888.
7. Schiller, F., Neligan, G. en Budtz-Olsen, O. E. (1948): Lancet. **2**, 842.
8. Walker, W. (1955): *Ibid.*, 749.

STERILIZED MILK AND MILK DRINKS

Two years ago Dr. J. A. Richter called attention at the South African Medical Congress to the introduction at Port Elizabeth of the production and marketing of sterilized milk; and the publication of Dr. Richter's[1] paper was accompanied by editorial comment[2] in this *Journal*. It is now generally accepted that milk supplies ought to be safeguarded by heat treatment to remove the potential threat of milk-borne disease, which dairy control has not otherwise succeeded in eliminating. Heat treatment is compulsory in certain parts of the world but in South Africa only in Cape Town, and much raw milk is still consumed in this country.

The prevailing method of heat treatment is pasteurization at as low a temperature as will in the time of operation kill the infection of ordinary milk-borne diseases. The extent to which pasteurization prolongs the time the milk can be 'kept' is very limited. Milk sterilization, which is carried out at a much higher temperature, in crown-corked bottles, kills all the germs in the milk so that it can be kept for practically an indefinite time without refrigeration.

The 'keeping' qualities of unopened bottles of sterilized milk are extremely convenient for the sale of milk over the counter, and for households that are not

equipped with refrigerators and, although pasteurization is still the process of choice for most purposes, sterilization has an important field of its own. Indeed Dr. Richter said in her paper that with one year's production of sterilized milk the consumption of milk in Port Elizabeth had increased by 10% at least. It has recently been stated that now that sterilized milk has been on sale there for 3 years 35% of all the milk sold in Port Elizabeth is consumed as sterilized milk.

One form of sterilized milk of considerable health interest is the flavoured milk-drink resembling a 'milk-shake'. Drinks, of course, will stand on their own merits as beverages, not as foods, and the various 'cool drinks' on the market will always appeal to a section, at all ages, of those who prefer them to cold water. But the fact remains that a great deal of money is spent on 'cool drinks' by people who would be better advised to buy something nutritious; and for these people milk drinks, not necessarily made with 'whole' milk, offer the advantage of being pleasing to the eye and the palate and at the same time a really valuable addition to the diet.

It has now been announced that a new factory has been completed in Johannesburg for the production of sterilized whole milk and sterilized flavoured milk as well as pasteurized milk.

1. Richter, J. A. (1954): S. Afr. Med. J., **28,** 762.
2. Editorial (1954): *Ibid.*, **28,** 757.