HAEMOPHILIA

Prof Julius Ogeng'o

University of Nairobi

E-mail : jogengo@uonbi.ac.ke

HAEMOPHILIA:

Potentially Lethal Bleeding

BY DR. J. A. OGENG'O

When boys bleed to death following circumcision; when people bleed excessively after a simple tooth extraction; when trivial bruises never appear to stop bleeding; when calf muscles seem to hurt after along walk; when gums tend to bleed unnecessarily; when joints lock in abnormal positions forever; when the boys in a family do not seem to survive beyond adolescence; when a young man is warned against marrying girls from a particular family.....

Haemophilia may be the reason!

Tom Okelle would never have married more than one wife if Haemophilia was not a problem. His first wife Akoth brought into the family this bad blood that will not clot. Akoth's first son died 24 hours after traditional circumcision due to excessive bleeding. Dick was Akoth's third son. He was crippled because of bleeding in the joints. This is when Tom was prevailed upon to marry a second wife. He was never man enough for more than one wife. In fact, even Ndunge, the concubine, was already giving him ulcers.

Anne Ngotho, was good. No bad blood at all. Healthy children, brilliant in school - but Kikuyu all the same. So the clan won't accept her. Children in the Kikuyu culture belong to the mother. In any case, they were girls.

Dr. Ogeng'o: "The diagnosis of haemophilia is relatively straight forward. The patient is almost certain to be male, and frequently a family history is present"

Third wife - Angela Mango was the most acceptable. Culturally compatible, normal sons with no bad blood. But Tom is then 50 years and the children are young. Last year the poor man went hypertensive and down with depression. Why? He cannot cope with the stress of three wives and fifteen children, some crippled. One of the wives not culturally acceptable in his clan.

This is how I got to talk to Tom at length about the causes of hypertension and depression. The moods are very low, the blood pressure is very high. "Very bad blood, brought in by Akoth my first wife," Tom said severally during our one hour chat. Perhaps all this would not have happened if Haemophilia was not there.

Haemophilia is a disorder in which blood does not clot normally, thus excessive bleeding. It is inherited. One of the factors (Factor VIII) required for the blood to clot is inadequate in blood. It is common, occurring 1 in 10,000 of the population, where studies have been done.

Females do not suffer the disease. They can only be carriers. They transmit the disorder to one half of their sons, and the "carrier state" to one half of their daughters. If however a man who has the disorder survived to adulthood, and married a "carrier lady", then half of the daughters would have double the dose of the bad blood, and may bleed to death, and half the boys would suffer. Do remember however that these are theoretical statistical expectations. One could have all normal sons and daughters; or all abnormal children.

The most obvious feature is bleeding. The only repri-he onset of the bleeding is characteristically delayed for several hours or days after injury or surgery. It may however persist for several days or weeks. In milder cases, bleeding does not occur spontaneously but injury or surgery may cause excessive bleeding.

Bleeding into the joints does occur spontaneously and cause pain, which may be severe, with swelling, warmth and muscle stiffness. With appropriate treatment, the problem in the joint settles in a few days, as the blood is reabsorbed. However, repeated episodes cause damage to the joint, with wasting of related muscles, leading to deformity and crippling. The knee joints are most frequently affected but ankles, hips shoulder, elbows and wrists are also involved.

Bleeding into muscles and other soft tissues is frequent and potentially disabling. When the bleeding occurs into a hollow organ, blood may come out of the orifices. For example one could vomit blood pass, bloody urine and/or stools or cough blood. This can be massive and horrifying. Indeed, any organ in the body may be the site of bleeding, often with lethal effects. Bleeding in the throat, in the brain, around the heart, lungs etc. can cause death due to pressure effects.

Frequently large clots may form in totally unexpected sites and cause untold diagnostic problems. Such episodes may be associated with fever and anaemia.

The diagnosis of haemophilia is relatively straight forward. The patient is almost certain to be male, and frequently a family history is present, and the clinical picture tends to be characteristic. Laboratory blood tests to assay the level of factor VIII are usually confirmatory. When X-rays, ultrasound, CT scans, urine tests and other tests of organ function are being done, it is usually to assess the effect of the disease.

Not all bleeding is due to haemophilia. Other causes include:

- Other minor bleeding disorders that are inherited.
- Vitamin deficiency especially C and K
- Cancer of the blood (Leukaemia)
- Bone marrow deficiencies due to various diseases
- Liver disease, infections etc
- Kidney diseases; snake bites etc
- Drugs, for example, Aspirin, Quinine etc

Accordingly, anybody presenting with features of excessive bleeding should be investigated thoroughly to exclude other causes and confirm haemophilia to be the cause of the bleeding.

Following confirmation of the diagnosis, the patients, their caretakers, fiends and parents should all participate in the treatment. Children should be reared in as normal a way as is compatible with physical safety. Watch out as a parent/guardian - emotional crippling can be as serious as that resulting from the bleeding. Victims of haemophilia are encouraged to participate in sports such as swimming or golf, and to avoid those involving body contact. A vocation should be sought which is not inherently dangerous. The victims should aim at becoming self-supporting adults capable of looking after themselves and living a full life.

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When the bleeding episode occurs, the aim of treatment is to correct the fluid/blood lost and stop the bleeding. Whole blood is usually transfused and/or clotting Factor VIII concentrates given. It is possible in fact to calculate exactly the amount of Factor VIII required. This may then be given at regular well worked out intervals.

Actively swollen joints are usually immobilized and local chilling with ice packs instituted in the early states. When the bleeding has stopped, active movement without weight bearing is good in order to prevent chronic limitation of movement. Careful physiotherapy is definitely useful.

For minor bleeding occurring at accessible sites, local measures such as cold and gentle pressure may be helpful.

Although the life expectancy of the haemophiliacs has been greatly prolonged by ability to replace the deficient Factor VIII, death as a complication of bleeding still occurs quite frequently. However, the actual outcome depends on the severity of the ill ness.

Patients with milder forms of haemophilia usually live a virtually normal life but may have severe bleeding after dental extraction, injury or surgery. Patients with severe disease may require frequent hospital admission and may develop crippling joint deformities. Stroke does occasionally occur, and anaemia is often a problem.

Each one of us can contribute to ease the burden of haemophilia on the patient and the community, in one or the other of the following ways:

- If you notice abnormal bleeding especially following trivial injuries, suspect a bleeding disorder and seek a doctor's opinion. This way "haemophiliac families" will be identified.
- Then avoid injections, and/or games that may cause bruising, scratching and do not accept surgery however minor unless the doctor assures you.
- If you know a haemophiliac or you are one, always volunteer this information to the doctor, whenever you have a chance.
- Once a diagnosis of haemophilia has been made, let the doctor educate you and/or your relatives and friends on the dos and don'ts of the disease. Try to learn if your facilities allow, how to give blood at home.
- Maintain regular contact with an experienced doctor so that whenever hospital treatment is required, it is not delayed unnecessarily.
- Avoid where/when known, intermarriage between members from families with a known history of haemophilia.

In these and other practical ways, many patients with severe disease may live productive lives and only a few become chronic invalids.

This so called bad blood need not kill or cause hypertension. Practical treatment and care is within reach. Alert your doctor now! ❑