We have performed platelet aggregation studies on two patients with FEVR—a 7-year-old boy with total blindness and his 14-year-old female cousin, who is mildly affected. The boy’s total blindness at 3 months of age has been described elsewhere. Neither subject had been exposed to any antiplatelet agents within the two weeks prior to the study. Coagulation studies were conducted and the results compared with the normal ranges of values in our laboratory. Platelet aggregation to added arachidonic acid (1.5 mmol/l), adrenaline (5 μmol/l), collagen (1-0 mg/l), ristocetin (1.5 g/l, 1.2 g/l, and 0.5 g/l) and adenosine diphosphate (ADP) (1 and 10 μmol/l) was within normal limits in each patient as compared with that of healthy controls. Platelet counts and morphology were normal, as were prothrombin times, partial thromboplastin times, and fibrinogen levels. Factor VIII and von Willebrand’s factor were also within the normal range of each patient. The only abnormality noticed was the boy’s bleeding time of 13 minutes (normal range 2–9 minutes) and his lack of adenosine triphosphate (ATP) secretion as measured by the luciferin-luciferase method using a lumi-aggregometer. This finding is consistent with storage pool disease. The cousin’s bleeding time and ATP secretion were normal.

Although we found no abnormality of platelet aggregation in response to arachidonic acid in either patient, the prolonged bleeding time and storage pool disease in the boy may have aggravated his condition, resulting in his total blindness, which brought him to medical attention originally. An additional feature of his history may be significant. Shortly after his birth his mother reports being treated with high doses of aspirin for endometriosis while breast feeding him. The combination of FEVR, his storage pool disease, and aspirin exposure may explain his severe clinical course. While Chaudhuri et al. ’s report of platelet aggregation defects in response to arachidonic acid has yet to be verified in other FEVR patients, this case supports the contention that platelet abnormalities may contribute to the severity of FEVR.

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References

Call it lentectomy

Sir, Congratulations to Drs Grossman and Peyman on the report of their very successful application of this procedure. Following Dr Peyman’s pioneering work in this area we adopted his procedure and have been similarly gratified with a low rate of complications and very successful results.

Dr Peyman has been a persistent advocate of the term ‘pars plicata’, which is anatomically correct. Now if we can just get him to call it a lentectomy, then he will have it 100% proper. (Ectopia lentis is another condition which is very well treated by his procedure.)

PAUL E ROMANO
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References

Sir, I appreciate the kind comments of Dr Romano on our paper. His suggestion is correct, and we will use the proper terminology.

GHOLAM A PEYMAN
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Eye protection for welders

Sir, Mr G P H Brittain emphasises the need for adequate eye protection at all times for amateur welding especially using the MIG welder (metal arc inert gas welder). Fortunately the two patients reported on by Mr Brittain had a full visual recovery. We reported a case of foveal injury by an amateur welder which produced permanent visual loss, further emphasising the potential seriousness of the injury that can occur.

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References

Obituary

Professor T Krwawicz

Professor Tadeusz Krwawicz, who died in September 1988 at the age of 78 years, was born in Lwow in 1910.

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