Screening for haemorrhagic disorders in paediatric patients by means of a questionnaire

I. Music; M. Novak; B. Acham-Roschitz; W. Muntean
Department of Paediatric and Adolescent Medicine, Medical University of Graz, Austria

Keywords
Von Willebrand disease

Summary
Aim: In children, screening for haemorrhagic disorders is further complicated by the fact that infants and young children with mild disease in many cases most likely will not have a significant history of easy bruising or bleeding making the efficacy of a questionnaire even more questionable. Patients, methods: We compared the questionnaires of a group of 88 children in whom a haemorrhagic disorder was ruled out by rigorous laboratory investigation to a group of 38 children with mild von Willebrand disease (VWD). Questionnaires about child, mother and father were obtained prior to the laboratory diagnosis on the occasion of routine preoperative screening. Results: 23/38 children with mild VWD showed at least one positive question in the questionnaire, while 21/88 without laboratory signs showed at least one positive question. There was a trend to more specific symptoms in older children. Three or more positive questions were found only in VWD patients, but only in a few of the control group. The question about menstrual bleeding in mothers did not differ significantly. Sensitivity of the questionnaire for a hemostatic disorder was 0.60, while specificity was 0.76. The negative predictive value was 0.82, but the positive predictive value was only 0.52. Conclusions: Our small study shows, that a questionnaire yields good results to exclude a haemostatic disorder, but is not a sensitive tool to identify such a disorder.

Material and methods
88 children, in whom a haemorrhagic disorder was ruled out by extensive laboratory investigation where compared to 38 children with mild VWD. The 88 children were recruited from a study comparing PFA-100 and aPTT in preoperative screening (12) and were investigated because of a minimal prolonged PTT and/or prolonged closure times in the PFA-100, but no specific haemorrhagic defect could be detected. The mild VWD-patients were detected because of prolonged PTT and/or PFA-100 closure times, VWF:Ag and VWF: RCoF ranged from 30–50%.

Questionnaires comprised three parts about child, mother, and father with nine questions each. Only yes/no answers were possible. Questionnaires were obtained prior to the laboratory work up on the occasion of routine preoperative screening. They were done by the parents alone while waiting and checked by young house staff trainees not especially trained in haemostatic disorders.

Statistical analysis were performed with the aid of SPSS 15.0 software (SPSS Inc., Chicago, Illinois, USA). Frequencies and percentages were analyzed by cross tabulation and the interrelationship between positive or negative questionnaires, and presence or absence of coagulopathy was tested by Fisher’s exact chi-square test. P-values < 0.05 were considered as statistically significant. Evaluating the quality of the used questionnaire as a diagnostic instrument for coagulopathy, a 2 by 2 table was performed to calculate the sens-

Preoperative screening for haemorrhagic disorders is an endless, but not very successful story. Poor results are obtained with laboratory screening alone. Questionnaires may be helpful according to a consensus. However, the efficacy of questionnaires to detect minor haemorrhagic disorders is not proven.

In children, screening for haemorrhagic disorders is further complicated by the fact that infants and young children with mild disease in many cases most likely will not have a significant history of easy bruising or bleeding. This fact makes the efficacy of a questionnaire even more questionable. There are ten studies about preoperative screening in children to prevent nonsurgical bleeding (1–10). They all agree that questionnaires should be used.

Questionnaires reduce the unnecessary use of laboratory tests.

Studies are less clear about sensitivity. Since only about 1–3 percent of children experience nonsurgical bleeding (11), only few children were available to investigate the efficacy of screening by questionnaire (8, 9) or clotting tests. In one study, only 11 of 27 children with nonsurgical bleeding had a positive questionnaire and in another study, only 4 of 22 children in whom a clotting disorder had been identified (8, 9).

We tried a different approach to investigate the sensitivity of questionnaires: We compared the questionnaires of children in whom a haemorrhagic disorder was ruled out by rigorous laboratory investigation to children with mild von Willebrand disease (VWD).
Results

When one positive question in one of the three parts was taken as positive in the sense of haemorrhagic disorder, 61% of VWD patients were detected; but 24% of questionnaires of the group of laboratory negatively tested children were also positive (Fig. 1a).

When two questions had to be positive in one of the three parts, 42% of VWD patients were detected, the positive results of questionnaires in the laboratory negative group decreased to 10% (Fig. 1b).

When three answers had to be positive only 8% of the laboratory negative had a positive result in the questionnaire, but only 26% of the VWD patients were detected (Fig. 1c).

Further analysis showed no significant differences in symptoms: Only bleeding during surgery and bleeding from unusual sites differed between families with VWD and the laboratory negatives. No differences were found for mucosal bleeding or cutaneous haematomas (Fig. 2). Very surprisingly, strong menstrual bleeding was more often checked by mothers of healthy children than by mothers of VWD patients (Fig. 3).

Sensitivity to detect a haemorrhagic disorder was 0.60, specificity 0.76. The negative predictive value of a questionnaire with no positive answer was 0.82, while the positive predictive value was only 0.52.

Discussion

Our study shows that a questionnaire is helpful to rule out a haemorrhagic disorder in children prior to first surgical procedure. It has to be emphasized that our study was done in an strict routine setting: Parents had to fill in the questionnaire without help, and evaluation of the questionnaire was done by young house staff. Of course, this is not the ideal to get best results with a questionnaire,
but that reflects the conditions prior to most routine surgical procedures. Very likely, results would be better, if an experienced haematologist would do the questioning, but this is rather unrealistic when surgery has to be done.

Therefore, our finding that under routine conditions the negative predictive value was 0.82 is encouraging. However, the positive predictive value was low. That means, that similar to screening by PTT or PFA-100 (12), many healthy children will have an unnecessary laboratory investigation. This will delay the planned procedure even with the use of a questionnaire.

**Conclusion**

Our small study shows that a questionnaire yields good results to exclude a haemostatic disorder, but it is not a sensitive tool to identify such a disorder. However, our study has a strong limitation: Patients with mild VWD were defined by laboratory means only and no patient bled during a surgical procedure since they all were treated with DDAVP. So, in some of these patients a negative questionnaire may be entirely correct.

**Conflict of interest**

All authors declare that there is no conflict of interest.

**References**