# PERIOPERATIVE MANAGEMENT OF CHILDREN WITH VON WILLEBRAND DISEASE AND TONSILLAR HYPERTROPHY IN ENT DEPARTMENT OF CHILDREN HOSPITAL IN WARSAW MEDICAL UNIVERSITY

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Summary

**Introduction.** The tonsillar hypertrophy are the most common children's disease. The treatment of choice is surgery. Regardless of surgery procedure perioperative bleeding is still present. The problem with homeostasis occurs in 0.5-2 percent of cases due to bleeding disorders. The most common inherited coagulopaties is von Willebrand disease (vWD). vWD commonly presents with epistaxis, excessive bleeding following soft tissue trauma, ecchymosis. Three type of vWD have been described. The treatment of hemorrhage in patients with vWD is i.v. infusion of Desmopressin (Minirin), concentrate containing factor VIII/vWF or antifibrinolytic drugs.

**Material and methods.** From January 2008 to December 2011 in the Department of Pediatric Otholaryngology of Medical University of Warsaw we treated by surgery 16 children with von Willebrand disease because of the tonsillar hypertrophy.

**Results.** 8 children with vWD and tonsillar hypertrophy underwent adenoidectomy and 8 adenoidotonsillotomy. Von Willebrand disease has been recognized because of clinical symptoms and blood tests in preoperative time. We treated by surgery 14 children with type 1 and 2 children with type 2 of vWD. 14 children has received Desmopressin (Minirin) intravenously and 2 children has received factor VIII-vWF concentrate (HaemateP).

8 children has received etamsylat (Cyclonamina), all 16 children has received as antifibrinolytic therapy tranexamicum acidum (Exacyl).

**Conclusions.** Adenoidectomy with/or without tonsillotomy in children with von Willebrand disease seems to be safe procedure by using adequate treatment of haemostasis.

Key words: adenoidectomy, tonsillotomy, children, von Willebrand disease

## INTRODUCTION

The tonsillar hypertrophy are the most common children's disease. The treatment of choice is surgery by adenoidectomy with/or without tonsillotomy. Regardless of surgery procedure perioperative bleeding is still present, but normally is stopped spontaneously. In 0.5-2 percent of cases we have a problem with homeostasis and some special treatment using haemostatic drugs has to be used. In 0.04 percent of patients the compatibility blood has to be transfused. The cause of severe hemorrhage is perioperative trauma of soft tissues in naso- or oropharynx, not complete excision of tonsils and haemorrhagic or vascular diathesis.

The most common inherited bleeding disorder is von Willebrand disease (vWD). 1% of general population has insufficiency or dysfunction of von Willebrand factor (vWF). Most of those deficiencies are clinically asymptomatic. Only in 1 of 10 cases of vWD the abnormal bleeding is observed. vWD is inherited in an autosomal -dominant often then autosomal-recessive pattern. vWF is encoded by a gen on chromosome 12 and synthesized in vascular endothelial cells and megacariocytes. The vWF has two haemostatic functions: preventing degradation of factor VIII and mediates platelet adhesion to damaged endothelium.

vWD commonly presents with epistaxis, excessive bleeding following soft tissue trauma, ecchymosis. Three type of vWD have been described. The most common is type 1 (75% of all) in them the bleeding is very mild, in type 2 bleeding is more excessive and in type 3 most severe due to absent of vWF. In type 1 the coagulopathy is characterized by partial quantitative vWF deficiency and in type 2 by qualitative defect in vWF. The treatment

of hemorrhage in patients with vWD is i.v. infusion of Desmopressin (Minirin), concentrate containing factor VIII/vWF or antifibrinolytic drugs. The knowledge of the type of vWD is very important in preoperative care to plan surgery procedures or substitute treatment.

## AIM

The aim of this study was to asses perioperative management of adenoidectomy or adenotonsillotomy in children with von Willebrand disease.

## MATERIAL AND METHODS

From January 2008 to December 2011 in the Department of Pediatric Otholaryngology of Medical University of Warsaw we treated by surgery 16 children with von Willebrand disease. All of them has tonsillar hypertrophy. It was 5 girls and 11 boys in age from 3 to 12 year. We analyzed preoperative laboratory tests, surgery technique, perioperative treatment of haemostasis and postoperative complications.

## RESULTS

All children with tonsillar hypertrophy and von Willebrand disease has been treated by surgery using La Force adenotome or Becman's knife and tonsillotome. 8 children underwent adenoidectomy and 8 adenoidotonsillotomy. At the same time in 3 children lower level of XII factor has been diagnosed and in 1 child spherocytosis. Von Willebrand disease has been recognized because of clinical symptoms and blood tests in preoperative time. The activated partial thromboplastin time (aPTT) was from 30.2 to 61,8 seconds and was extended in 9 children. Activity of ristocetin cofactor of vWF (vWF:RCo) was from 36.12 to 54.2 and was decreased in all children, the level of antigen vWF (vWF:Ag) was from 38,4-52,8 and was decreased in 15 children. The activity of factor VIII was from 47,3 to 92.8 and was decreased in 1 child. The plates level was normal in all children, as well as the INR factor (0.98 to 1,23). 10 children has blood group "0", 4 children "A" and 2 children "B".

We treated by surgery 14 children with type 1 and 2 children with type 2 of vWD. Just 60-90 minutes before surgery 14 children has received Desmopressin (Minirin) in one dose 0.3-0.4 mcg/kg of the body weight intravenously. 2 children has received factor VIII-vWF concentrate (HaemateP). One child has received it in one dose, other in few doses. First dose has been given before surgery, next after 12 hours and next doses every 24 hours in the following 6 days. 8 children has received etamsylat (Cyclonamina) in dose 9 mg/kg every 8 hours in the next 9 days from start intravenously, then orally. All 16 children has received as antifibrinolytic therapy tranexamicum acidum (Exacyl) in dose 20 mg/kg every 6-8 hours in 8 days with first dose the day before surgery.

As a haemostatic management during surgery the tampon into the nasopharynx for 5-7 minutes has been put and the rest of palatine tonsils was pressed by gausepad. The bleeding was no more than 100 ml. In 15 children we has not observed bleeding in postoperative time. In 1 child small bleeding has been occurred in 1 day after surgery. It has been stopped by additional substitution of the Haemate P to Desmopressin. The average hospitalization was 6.3 days.

## DISCUSSION

Von Willebrand disease is quiet popular in general population, and almost has been diagnosed by chance in asymptomatic children who underwent blood tests before surgery procedures. Very careful clinical

Table 1. Perioperative management of children with tonsillar hypertrophy and vWD.

| Patient | Age | Days in hospital | Type of vWD | Type of surgery procerures | Drugs |
|---|---|---|---|---|---|
| D.P. | 12 | 8 | 2 | adenotonsillotomy | Haemate P |
| Ł.R | 5 | 6 | 1 | adenotonsillotomy | Desmopressin |
| K.W. | 6 | 7 | 1 | adenotonsillotomy | Desmopressin |
| D.A. | 4 | 8 | 1 | Adenotomy | Desmopressin |
| S.M. | 8 | 7 | 1 | adenotonsillotomy | Desmopressin |
| K.Z. | 6 | 7 | 1 | adenotonsillotomy | Desmopressin |
| D.R-B | 6 | 7 | 1 | adenotomy | Desmopressin |
| K.S. | 3 | 5 | 1 | adenotonsillotomy | Desmopressin |
| K.R | 5 | 6 | 1 | adenotomy | Desmopressin |
| A.J. | 4 | 3 | 1 | adenotomy | Desmopressin |
| W.O. | 7 | 6 | 1 | adenotomy | Desmopressin |
| K.C. | 9 | 7 | 1 | adenotomy | Desmopressin |
| G.Cz. | 8 | 7 | 1 | adenotomy | Desmopressin |
| F.W. | 3 | 6 | 1 | adenotonsillotomy | Desmopressin |
| Z.O. | 8 | 7 | 2 | adenotonsillotomy | Haemate P |
| P.P | 4 | 5 | 1 | adenotomy | Desmopressin |

investigation of children and it's family history of epistaxis, abnormal bleeding and bruising has to be done. If the investigations suggested the coagulopathy special blood test should be done before surgery. Excessive activated partial thromboplastin time (aPTT), decreased activity of ristocetin cofactor of vWF, lower level of antigen vWF and decreased activity of coagulation factor VIII describe von Willebrand disease type 1. The treatment of choice is intravenous infusion of Desmopressin (DDAVP). It's caused transient increase vWF and factor VIII in plasma. This infusion is not effective in type 3 and in most patients with type 2. The desmopressin test have to be done in all patients before start to asses response to this treatment. The dose of DDAVP is 0.3 mcg/kg of body weight diluted in 50-100 ml of isotonic saline solution in i.v. infusion over 30 minutes 90 minutes before surgery. The treatment of DDAVP can be done no more than 3-5 days due to exhaustion of endothelial storage of vWF. Type 2 of vWD is composed of few subtype (2A, 2B, 2N, 2M) with various defects of the vWF activity. The study of multimer patterns of vWF and its reaction on thrombocytes and collagen have to be done to diagnosed subtype of vWD. In type 2N, 2A, 2M where the levels of normally functioning vWF in plasma is mildly or moderately decreased, the intravenous infusion of DDAVP is effective. The rest patients can be treated with concentrates containing vWF/factor VIII replacement therapy. We used product Haemate P. Haemate P can be used as well in patients with vWD who were unresponsive to DDAVP, who may have contradictions for use DDAVP or when the treatment have to be done over 3-5 days. Additionally tranexamicum acidum (Exacyl) can be given orally or intravenously at a dose 20 mg/kg of body weight every 8 to 12 hours except the patients with haematuria. In very small postoperative haemorrhage. Exacyl can be used in monotherapy (14).

## CONCLUSIONS

Adenoidectomy with/or without tonsillotomy in children with von Willebrand disease seems to be safe procedure by using adequate treatment of haemostasis. □

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