Thieme

Perioperative Management of Coagulation Disorders in Ophthalmic Surgery

Perioperatives Management bei Gerinnungsstörungen in der Ophthalmochirurgie

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ABSTRACT

Disorders of blood coagulation can lead to manifest spontaneous bleeding and an increased risk of bleeding during surgical procedures and interventions. Pathophysiologically, a distinction can be made between defects in primary haemostasis, which lead to impaired platelet adhesion and platelet aggregation, and disorders of secondary (plasmatic) haemostasis, which are characterised by impaired fibrin formation or fibrin stabilisation. Aetiologically, a distinction can be made between rare genetically-determined hereditary defects and common acquired coagulation disorders, which may be based on different pathomechanisms. This overview is intended to provide ophthalmic surgeons with a basis for the perioperative management of patients with genetically determined coagulation disorders undergoing ophthalmic surgery. As there are no specific recommendations in this regard, the recommendations are based on the procedure for other surgical interventions, taking into account the specific bleeding risk associated with ophthalmic surgery.

ZUSAMMENFASSUNG

Störungen der Blutgerinnung können zu spontanen Blutungsmanifestationen und einem erhöhten Blutungsrisiko bei operativen Eingriffen und Interventionen führen. Pathophysiologisch lassen sich Defekte der primären Hämostase, die zu einer gestörten Thrombozytenadhäsion und Thrombozytenaggregation führen, sowie Störungen der sekundären (plasmatischen) Hämostase, die durch eine Beeinträchtigung der Fibrinbildung oder Fibrinstabilisierung charakterisiert sind, unterscheiden. Ätiologisch kann man seltene genetisch determinierte hereditäre Defekte und häufige erworbene Gerinnungsstörungen unterscheiden, denen unterschiedliche Pathomechanismen zugrunde liegen können. Mit dieser Übersicht soll Ophthalmochirurgen eine Grundlage zum perioperativen Management von Patienten mit genetisch determinierten Gerinnungsstörungen gegeben werden, die sich einem ophthalmochirurgischen Eingriff unterziehen müssen. Da diesbezüglich keine spezifischen Empfehlungen existieren, beruhen die Empfehlungen auf dem Vorgehen bei sonstigen operativen Eingriffen, wobei das spezifische Blutungsrisiko im Rahmen der Ophthalmochirurgie berücksichtigt wird.

Introduction

Coagulation disorders may cause spontaneous bleeding and increased bleeding risk during surgical operations and interventions. A pathophysiological distinction can be drawn between primary haemostasis disorders leading to impaired platelet adhesion and platelet aggregation on the one hand, and secondary (plasmatic) haemostasis disorders exhibiting fibrin formation or stabilisation impairment on the other. There is also an aetiological distinction between rare genetically determined hereditary defects

and common acquired coagulation defects caused by a wide variety of possible pathological mechanisms.

This review aims to provide ophthalmic surgeons with a basis for perioperative management in ophthalmic surgery patients with genetically determined coagulation disorders. Recommendations specific to this topic do not exist; generalised recommendations from other operations are used instead, taking the specific risks of bleeding in ophthalmic surgery into account.

This contribution will not cover perioperative management of antithrombotic medication, as we have already published recommendations on this topic [1–5].

Blood Coagulation – the Fundamentals

Blood coagulation is a complex process; its purpose is to stem bleeding due to vascular injury, ensuring vascular integrity. Coagulation is a complex process as it involves blood as a moving fluid but needs to remain local to the site of the lesion and cease altogether after effective haemostasis. A reduction in coagulation may lead to an increased tendency to bleed, whereas excessive blood coagulation may cause thrombotic complications [6].

Ensuring effective haemostasis requires precise interaction between the many components of the coagulation system with its extreme complexity and many regulating mechanisms. Apart from haemostasis, the coagulation system includes antithrombotic mechanisms to halt an activated coagulation process and deactivate the activated coagulation (protein C/protein S system), limit the coagulation process to the vascular lesion site (antithrombin), and dissolve the clot that has formed (fibrinolysis).

Coagulation comprises primary and secondary (plasmatic) haemostasis in addition to antithrombotic mechanisms. Importantly, these processes undergo complex interplay and occur almost simultaneously in vivo although they are viewed in isolation to aid understanding.

Primary haemostasis

Primary haemostasis refers to the process of platelet adhesion and aggregation. This is triggered by the endothelial lesion caused by vascular trauma. A circulating adhesive protein, von Willebrand factor (vWF), binds with high affinity to the exposed subendothelial collagen and undergoes a conformational change. This allows platelets to bind to vWF via a specific glycoprotein (GP) receptor complex, GP Ib-V–IX, leading to initial platelet adhesion; interaction between other platelet receptors, such as collagen receptors GP VI and GP Ia-IIa, and the subendothelial matrix reinforces this initially weak binding. This adhesion process increases platelet activation, which includes increased expression, clustering, and activation of platelet aggregation receptor GP IIb-IIIa; these receptors enable platelets to bind to each other, a process mediated by fibrinogen or vWF [6].

Secondary (plasmatic) haemostasis

Secondary (plasmatic) haemostasis refers to the fibrin formation and stabilisation process. This is also a complex process involving many plasmatic coagulation factors with many inhibitors and regulators regulating this process. Like primary haemostasis, an endothelial lesion also triggers secondary haemostasis; exposure of subendothelial tissue factor (TF), formerly known as tissue thromboplastin, to the circulating blood plays a decisive role here. TF binds to circulating coagulation factor VII, causing factor VII activation into factor VIIa. Other coagulation factors and cofactors contribute to forming thrombin from its precursor, prothrombin. Thrombin activates the platelets, creating a catalytic platform on the platelets for large amounts of thrombin to be formed under the influence of other coagulation factors – the characteristic thrombin burst. Thrombin catalyses fibrin formation from its precursor, fibrinogen, while also cross-linking fibrin molecules via fibrin-stabilising factor, factor XIII [6, 7].

Antithrombotic mechanisms serve to place limits on the duration and location of the activated coagulation process. The **protein C/S system** deactivates the activated coagulation process, thus limiting coagulation duration. **Antithrombin** restricts the coagulation process to the site of the vascular lesion in the moving blood by inhibiting activated coagulation factors carried away in the bloodstream. Defects in the protein C/S system and antithrombin do not lead to an increased tendency to bleed but play a crucial role in thrombosis risk. Endogenous **fibrinolysis** enables dissolution of fibrin clots; excessive fibrinolysis (hyperfibrinolysis) may lead to an increased tendency to bleed [6]. However, hyperfibrinolysis is usually caused by acquired causes that do not play a significant role in ophthalmology, so the present contribution will not be covering this aspect any further.

Coagulation Defects with Increased Bleeding Tendency

Primary haemostasis disorders

Primary haemostasis disorders can involve the platelets themselves (thrombocytopenia, platelet dysfunction) or von Willebrand factor (Von Willebrand disease).

Thrombocytopenia

Thrombocytopenia is characterised by a reduction in platelets below the normal range of 150,000–400,000/µl. However, pseudothrombocytopenia is an important consideration as it appears in around 0.1 % of blood samples; this is a clinically irrelevant laboratory artefact where anticoagulant ethylenediaminetetraacetic acid (EDTA) in the blood count tube causes platelet agglutination in vitro, which simulates thrombocytopenia. Reduced platelet counts in EDTA blood compared to normal values in an alternative collection medium (citrated blood, special tubes for platelet determination) indicate a pseudothrombocytopenia diagnosis. Affected individuals do not exhibit any increased tendency to bleed and therefore do not require treatment or bleeding prevention drugs during surgery.

"True" thrombocytopenia is almost always acquired; congenital forms are rare. Thrombocytopenia has a highly complex and multifaceted aetiology and pathogenesis. However, aetiological clarification of thrombocytopenia is extremely important as the treatment approach will depend on its aetiopathogenesis; whether the disorder involves reduced synthesis, increased consumption, or increased degradation of platelets plays a major role [8–10].

▶ **Table 1** Therapeutic measures for thrombocytopenia depending on platelet count and clinical situation (*pure thrombocytopenia, no additional platelet dysfunction).

Clinical situation	Platelet count*	Bleeding prevention and treatment
Any	> 100,000/µl	None
Surgical interventions, not high-risk	> 50,000/µl	None
	< 50,000/µl	Platelet concentrate (aim for platelet count at > 50,000/µl)
High-risk procedures	> 100,000/µl	None
	70,000–100,000/µl	Consider platelet concentrate
	< 70,000/µl	Platelet concentrate (aim for platelet counts at > 70,000–100,000/µl)
Severe bleeding	< 100,000/µl	Platelet concentrate (aim for platelet count at > 50,000–100,000/µl)

There are many differential diagnoses for thrombocytopenia: The most common cause in adults is immune thrombocytopenia (ITP) where antiplatelet antibodies destroy the platelets without sufficient new platelet production to compensate for the loss. Other common causes of thrombocytopenia include hepatic diseases, especially liver cirrhosis; pathogenetically, impaired thrombocytopoiesis and increased storage of platelets ("pooling") in a spleen enlarged due to portal hypertension (hypersplenism) play a key part in this. In addition, thrombocytopenia may occur due to conditions such as systemic haematological diseases and drug-induced bone marrow damage impairing the bone marrow's capability to synthesise platelets. Gestational thrombocytopenia occurs during pregnancy and is mainly due to a physiological increase in plasma volume; however, this form of thrombocytopenia is usually only mild and not clinically relevant. Finally, acute diseases especially such as disseminated intravascular coagulation (DIC) may lead to reduced platelet counts; these cases require treatment of the underlying disease as a decisive measure. In summary, thrombocytopenia is extremely complex in its aetiology and pathogenesis, and may require complex diagnostics to clarify [8–10].

Patients with thrombocytopenia may be at increased risk of perioperative bleeding. Considerations as to whether a particular case involves isolated thrombocytopenia or an additional platelet disorder are essential, as this can substantially increase the risk of bleeding in thrombocytopenia. Note also that concomitant thrombocytopenia makes platelet dysfunction difficult to identify [8–10].

Platelet counts of more than 50,000/µl are generally considered sufficient in operations (except high-risk procedures) if platelet function is undisturbed and no other coagulation disorders are present. Procedures presenting minimal risk of bleeding can usually be performed without problems, even with lower platelet counts. High-risk procedures, especially neurological operations, require higher platelet counts of 70,000–100,000/µl. Platelet counts should be raised to more than 100,000/µl by platelet transfusion in cases of substantial bleeding, especially with potential loss of vision [11]. ▶ **Table 1** below presents a perioperative transfusion strategy for thrombocytopenia.

Some ophthalmic operations potentially involving persistent visual impairment or loss may also be considered high-risk procedures; ophthalmic surgeons responding to a survey we carried out especially identified orbital surgery and fistulising glaucoma as high-risk procedures [1].

Platelet dysfunction

Platelet dysfunction (thrombocytopathy) encompasses a heterogeneous range of clinical conditions characterised by platelet function impairment. Congenital or hereditary thrombocytopathy is extremely rare; defects in platelet receptors (such as Glanzmann thrombasthenia) and storage pool defects [SPD] causing granular abnormalities in platelets) [12] are potential causes. In contrast, acquired platelet dysfunction is extremely common. Platelet function-inhibiting drugs are the most common explanation for acquired platelet dysfunction. Platelet inhibition may indeed be the intended effect in some drugs such as acetylsalicylic acid and thienopyridines (clopidogrel, prasugrel), but a host of other pharmaceuticals can also cause a disruption in platelet function as a side effect. Analgesics are a frequent cause, especially those containing aspirin and non-steroidal anti-inflammatory drugs (NSAIDs); however, many other pharmaceuticals such as selective serotonin reuptake inhibitors (SSRIs) and antibiotics are also potential candidates. Acquired platelet disorders also arise in liver cirrhosis (hepatic thrombocytopathy), end-stage renal failure (renal thrombocytopathy), and haematological diseases [13].

Thrombocytopenia is easily recognisable in blood counts, but routine diagnostics will not detect platelet dysfunction. Importantly, platelet function cannot be inferred from platelet counts; apart from that, results from both Quick value and aPTT remain unremarkable in isolated platelet dysfunction. Detection therefore requires specialised procedures, whereby aggregometry serves as the gold standard in diagnostics [14].

Critical review of drugs impairing platelet function is essential in patients exhibiting platelet dysfunction. Corresponding drugs may be paused perioperatively if necessary, while also taking the risk of serious thrombotic complications into consideration on discontinuing platelet function inhibitors. Preoperative suspension of platelet function inhibition usually requires interdisciplinary consultation between ophthalmic surgeons and attending cardiologists/angiologists.

Various options exist for preventing and treating bleeding in platelet dysfunction, depending on pathogenesis. More effective

▶ **Table 2** Therapeutic options for platelet dysfunction (thrombocytopathy).

Drug	Dosage	Mechanism of action
Antifibrinolytic	Tranexamic acid: 2–3 × 500–1,000 mg/d	Fibrinolysis inhibition
Desmopressin (DDAVP)	0.3–0.4 µg/kg bodyweight as a short infusion for approx. 30 min (NOTE: Tachyphylaxis possible with repeated application)	Release of von Willebrand factor and factor VIII from endogenous stores, prohaemostatic effect
Recombinant activated factor VII (rFVIIa)	100 µg/kg bodyweight, repeat administration if necessary	Prohaemostatic effect, massive thrombin generation
Platelet concentrate	Variable, initially 1–2 platelet concentrates at 24×10^{11} platelets	Replace dysfunctional with functional platelets

dialysis may temporarily improve platelet function in uraemic thrombocytopathy, thus reducing the risk of perioperative bleeding. Drugs used to manage mild platelet dysfunction include antifibrinolytics, especially tranexamic acid, and desmopressin (DDAVP), a vasopressin analogue. Recombinant activated factor VIIa may also be administered in some cases. A transfusion of platelet concentrates may also be required (▶ **Table 2**) [12, 13].

Von Willebrand disease

Von Willebrand disease (VWD) is the most common congenital/genetically determined coagulation disorder associated with bleeding risk; up to 600,000 people in Germany are affected according to estimates, although the number of unreported cases is likely to be high. This is because prolonged aPTT reveals only around 30% of those affected; the remaining cases require specific tests especially involving Willebrand factor (vWF) activity and concentration as well as factor VIII activity measurement. Specialised tests such as multimer analysis are used for classifying this heterogeneous disease rather than its diagnosis.

Autosomal inherited congenital VWD exhibits a reduction (type 1), a functional defect (type 2), or absence of VWD (type 3); this varies widely between individuals [15–17]. Acquired Von Willebrand disease accounts for approximately 10% of all cases and is pathophysiologically heterogeneous; the present contribution will not be covering this condition any further beyond reference to further literature [18].

Determining the appropriate prevention and treatment strategy for bleeding in patients with VWD is a complex process and depends on type and severity. The desmopressin (DDAVP) vasopressin analogue is the preferred treatment in mildly responsive forms; this releases vWF and factor VIII from endogenous stores. Coagulation factor concentrates containing vWF are used for substitution the absence of response to DDAVP and in severe manifestations of VWD. Antifibrinolytics may also be used as adjuvant treatment [15–17].

▶ **Table 3** below shows treatment options for VWD.

Secondary (plasmatic) haemostasis disorders

Plasmatic coagulation disorders exhibit deficiency, dysfunction, or absence of coagulation factors and are associated with reduced fibrin formation or stabilisation [19].

▶ **Table 3** Prevention and treatment of bleeding in Von Willebrand disease.

Type	Subtype	Bleeding prevention and treatment
Type 1	–	▪ Mild manifestation: Desmopressin (DDAVP) ▪ Severe manifestation: factor concentrate containing vWF
Type 2	2A	▪ Mild manifestation: Desmopressin (DDAVP) if response is sufficient, otherwise factor concentrate containing vWF. ▪ Severe manifestation: factor concentrate containing vWF
	2B	Factor concentrate containing vWF
	2 M	▪ Mild manifestation: Desmopressin (DDAVP) if response is sufficient, otherwise factor concentrate containing vWF. ▪ Severe manifestation: factor concentrate containing vWF
	2 N	Factor concentrate containing vWF
Type 3	–	Factor concentrate containing vWF

All hereditary plasmatic coagulation disorders are very rare. The most well-known form is haemophilia characterised by a reduction in factor VIII (haemophilia A) or factor IX activity (haemophilia B); X-chromosomal inheritance dictates that men are predominantly affected while women are carriers who may show moderate reduction in factor VIII or IX activity. Factor I (A), dysfibrinogenaemia, hypofibrinogenaemia, factor II, factor V, factor VII, factor XI (formerly known as haemophilia C), and factor XIII deficiency have autosomal inheritance patterns and are therefore gender-independent [19]. In contrast, common factor XII, prekallikrein, and high-molecular weight kininogen (HMWK) deficiencies are not generally associated with bleeding risk and can therefore be considered clinically irrelevant.

Acquired plasmatic coagulation defects are comparatively common and occur in conditions such as liver cirrhosis, vitamin K deficiency, and systemic amyloidosis. High blood loss (loss coagu-

▶ **Table 4** Prevention and treatment of bleeding in plasmatic coagulation disorders.

Coagulation defect	Minimal residual activity Before surgery	Bleeding prevention and treatment
Factor I deficiency (afibrinogenaemia, hypofibrinogenaemia, dysfibrinogenaemia	> 100 mg/dl	Fibrinogen concentrate
Factor II deficiency	?*	PPSB
Factor V deficiency	5 %	GFP (15–20 ml/kg bodyweight to achieve levels of 15–20 %)
Factor VII deficiency	Minor operations: 30–50 %, major operations: > 50 %	Factor VII concentrate
Factor VIII deficiency (Haemophilia A)	Minor operations: 25–40 %, major operations: > 50–80 %	DDAVP (subhaemophilia A, mild haemophilia A), factor VIII concentrate
Factor IX deficiency (Haemophilia B)		Factor IX concentrate
Factor X deficiency	?*	Factor X concentrate if available, otherwise PPSB
Factor XI deficiency	5 %	Factor XI concentrate if available, otherwise DDAVP and antifibrinolytics; GFP (20 ml/kg bodyweight to achieve levels of 20 %) on insufficient response
Factor XII deficiency	–	No increased tendency to bleed
Factor XIII deficiency	> 50 %	Factor XIII concentrate
HMWK deficiency	–	No increased tendency to bleed
Prekallikrein deficiency	–	No increased tendency to bleed

PPSB = prothrombin complex preparation, GFP = coagulation-active fresh plasma, rFVIIa = recombinant activated factor VII, DDAVP = desmopressin; *hardly any data available, case-by-case decision

lopathy), especially in inadequate volume replacement (dilution coagulopathy), and increased factor consumption (consumption coagulopathy) may also lead to complex plasmatic coagulation disorders [19].

Prolonged coagulation times from determining prothrombin times according to Quick (or Quick value decrease), activated partial thromboplastin time (aPTT) and/or thrombin times indicate plasmatic coagulation disorder. Abnormal findings in these coagulation tests require further clarification to establish a diagnosis, especially by determining individual factor activities [19].

Single factor concentrates, if available, are administered for substitution in factor deficiency to raise factor activity to the required level preoperatively; this depends on the type of coagulation defect present and, if applicable, the assumed risk of bleeding involved in the respective procedure. Dosing intervals for factor concentrates depend on the respective coagulation factor's half-life in the blood and vary according to the diverse types of coagulation defects. Prothrombin complex preparations (PPSB) can be used to replace vitamin K-dependent coagulation factors (II, VII, IX and X) in the absence of single factor concentrates. Fresh coagulation-active plasma (FFP) is applied in rare cases; this contains all the coagulation factors (and inhibitors). Desmopressin (DDAVP) can release factor VIII from endogenous stores and temporarily increase factor VIII activity in patients with mild haemophilia A [11, 19]. ▶ **Table 4** below shows therapeutic prevention and treatment options for bleeding in various plasmatic coagulation defects.

It is important to emphasise that very rare plasmatic coagulation defects do not have enough reliable data available for conclusive determination of an adequate perioperative bleeding prevention strategy. Decisions are usually made for each individual patient in these rare cases; the decision needs to consider not only the factor activity, but also the patient's individual bleeding tendency as well as the risk of bleeding involved in the respective surgical procedure. We have little in the way of data for ophthalmic surgery, so recommendations for other procedures need to be followed for orientation.

Ophthalmic Surgery: Recommendations for Preventing Bleeding in Patients with Coagulation Disorders

Risk stratification

Even without systematic data, we can assume that genetically determined, congenital, and acquired coagulation defects all increase perioperative bleeding risk during ophthalmic surgery. However, there is a lack of reliable data relevant to ophthalmic surgery due to the rarity of these disorders. It is important to bear in mind the specific risk of persistent partial or total loss of vision due to severe bleeding in ophthalmic surgery. ▶ **Table 5** below shows major ophthalmic operations classified by perioperative bleeding risk.

In the opinion of the authors, there is no need for routine preoperative diagnostics before ophthalmic surgery in procedures exhibiting low bleeding risk.

Current status is required for patients with a known coagulation defect before surgical interventions with an intermediate or

▶ **Table 5** Risk of bleeding in frequent ophthalmic operations [1, 2].

Low bleeding risk	Intermediate bleeding risk	High bleeding risk
▪ Cataract surgery ▪ Intravitreal injections (including implants) ▪ Laser operations ▪ Minor eyelid surgery ▪ Simple muscle surgery ▪ Corneal surgery	▪ Scleral indentation ▪ Vitrectomy ▪ Stent implants for glaucoma	▪ Extensive eyelid surgery ▪ Extensive orbital operations ▪ Major muscle surgery ▪ Valve implants for glaucoma ▪ Intraocular tumour surgery

▶ **Table 6** Bleeding prevention during ophthalmic surgery depending on perioperative bleeding risk and severity of a genetically determined coagulation defect.

	Low bleeding risk	Intermediate bleeding risk	High bleeding risk
Mild coagulation defect	No bleeding prevention	Low-risk prevention	High-risk prevention
Intermediate coagulation defect	Low-risk prevention	Low-risk prevention	High-risk prevention
Severe coagulation defect	Low-risk prevention	Low-risk prevention	High-risk prevention

high risk of bleeding or pronounced tendency to bleed, bearing in mind that coagulation findings may be subject to considerable fluctuation in the same patient such that old findings are often problematic. The following situations require increased vigilance:

- Thrombocytopenia with a previously known platelet count less than 50,000/µl
- Severe thrombocytopathy (such as Glanzmann's thrombasthenia)
- Von Willebrand disease with von Willebrand factor activity less than 50%
- Haemophilia A or B
- Factor VII or factor XIII deficiency with residual activity less than 50%
- Factor V or factor XI deficiency with residual activity less than 5%
- Hypofibrinogenaemia or dysfibrinogenaemia with fibrinogen levels less than 100 mg/dl

Overall, these are very rare situations; the number of required preoperative analyses in cases of known coagulation defects should be kept to a minimum. Cooperation with a haemostasiologist is recommended in such cases to determine appropriate perioperative management. As previously emphasised, the decision will often be made on a case-by-case basis for lack of reliable data on ophthalmic surgery.

Bleeding history is mainly used preoperatively to assess whether a coagulation defect or increased risk of bleeding is present in patients without a known coagulation defect. This history involves enquiring about bleeding events in the patient's personal and family history using a standardised questionnaire as applicable; these questionnaires are also commercially available. Negative bleeding history has been established to exhibit a high negative predictive value while 30–50% of patients with a conspicuous bleeding history also have a coagulation defect; however, our investigation did not reveal any conclusive association with increased perioperative bleeding risk in ophthalmic surgery, even in cases with conspicuous standardised bleeding histories.

The relevance of preoperative coagulation diagnostics is minor compared to bleeding history when identifying patients at increased risk of bleeding. Most common coagulation defects (Von Willebrand disease, platelet dysfunction) are not detected or are only partially detected in preoperative testing, which is one of the reasons for the low predictive value of routine diagnostic testing for perioperative bleeding risk. Routine determination of blood count, Quick value and aPTT may therefore be omitted if there is no evidence of any increased tendency to bleed in the patient history.

However, a specialised preoperative haemostaseological diagnosis should be performed to optimise perioperative management in patients with a conspicuous bleeding history undergoing ophthalmic operations associated with an intermediate or high risk of bleeding. This requires establishing a working relationship between ophthalmic surgery and haemostaseology.

Preoperative management

Specific recommendations do not yet exist for managing patients affected during ophthalmic surgery. This requires drawing on experience from other surgical disciplines that have also been incorporated into current guidelines [11]. Perioperative coagulation management always depends on the type and severity of the existing coagulation disorder coupled with the risk of bleeding during the respective operation.

Prohaemostatic medication for ophthalmic procedures should only be considered if a patient does not satisfy the limits required for surgery such that bleeding prevention drugs are not necessary in patients with mild coagulation defects that do satisfy these limits.

Specific bleeding prevention drugs are usually unnecessary in cases of mild to moderate genetically determined coagulation defects at an explicitly low risk of bleeding, but patients with severe coagulation disorders and/or moderate to high risk of bleeding will require bleeding prevention. Recommendations for general surgical interventions can be used for setting therapeutic goals in cases of low to medium bleeding risk, while neurosurgical interventions may be used in ophthalmic operations on the fundus or surgical interventions associated with a relevant risk of persistent visual impairment. ▶ **Table 6** below shows the basic procedure that may of course be modified in individual cases.

Conflict of Interest

Prof. Dr. Zeitz: Beratungs- und Vortragshonorare von AbbVie, Alexion, Bayer, Boehringer-Ingelheim, Novartis, Roche; Beratungshonorare von Immunocore, Omeicos, Oxular, SamChungDang, ViGeneron; Forschungsgelder von Bayer, Boehringer-Ingelheim, Novartis.

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