Chapter 7
Platelet Function Disorders

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ABSTRACT
Platelets play an important role in thrombosis and hemostasis. Moreover, platelet dysfunction due to congenital and acquired etiologies is also one of the most common causes of bleeding encountered in clinical practice. Mostly, platelet function disorders are deficiencies of glycoprotein mediators of adhesion and aggregation, whereas defects of primary receptors for stimuli include those of the P2Y12 ADP receptor. Studies on inherited defects of (1) secretion for storage organelles (dense and alpha-granules), (2) the platelet cytoskeleton, and (3) the generation of pro-coagulant activity have allowed for the identification of genes directly and/or indirectly controlling specific functional responses. This chapter will review recent advances in the molecular characterization of platelet function defects, the spectrum of clinical manifestations of these disorders and their management.

INTRODUCTION
Abnormality in platelet function causes bleeding in patient which is disturbance in hemostasis. The term hemostasis applies to a myriad of physiological processes that are involved in maintaining vascular integrity and keeping the blood in fluid form. Human platelets are multifunctional anucleated cells that play an important role in hemostasis. Here we will discuss the physiology of platelets in hemostasis and defects in platelet function.

Platelet Structure and Function
Platelets originate from the cytoplasm of bone marrow megakaryocyte (Figure 1). It lack genomic DNA but contain megakaryocyte-derived mRNA and the translational machinery needed for protein synthesis. Circulating platelets are discoid in shape, which dimensions of approximately 2-4 μm. Their shape and small size enables the platelets to be pushed to the edge of vessels, placing

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them in the optimum location to constantly survey the integrity of the vasculature. Platelets circulate in a concentration of 150,000–450,000 cells/mL. Of the total body platelets, about 70% stay in the circulation while the remaining 30% are continually but transiently sequestered in the spleen. Platelets remain in circulation for an average of 10 days (Kile, 2014). Most platelets are removed from the circulation by the spleen and liver after senescence, but a constant small fraction is continually removed through involvement in maintenance of vascular integrity (Kile, 2014).

On peripheral blood smears stained with Wright-Giemsa stain, platelets appear as small, granular staining cells with a rough membrane, and are normally present as 3-10 platelets per high-power oil-immersion field (Dunning, 2011). Despite their simple appearance on the peripheral blood smear, platelets have a complex structure (Figure 2). Platelets internal structure has been divided into four zones:

- Peripheral zone,
- Sol-gel zone,
- Organelle zone, and
- Membrane zone.

The peripheral zone includes the outer membranes and closely associated structures. The platelet has a surface-connected system of channels called the open canalicular system (OCS). The walls of the OCS are included in this zone. The OCS provides access to the interior of the platelet to plasma membranes, and an outlet channel for platelet products. The release of platelet products through the OCS after platelet activation is called “the release reaction”.

The membranes of the platelet are rich in platelet receptors, which determine its specific cellular identity. These receptors are constitutively expressed on the platelets and require conformational change during platelet activation to express receptor function. The major classes of receptors and their ligands are shown in Table 1.

The peripheral zone also includes membrane phospholipids (Kowata, 2014). Phospholipids are an important component of coagulation as they provide the surface upon which coagulation protein react.
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