

# Platelet Storage Pool Deficiency of $\alpha$ and $\delta$ Granules

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**A** 23-year-old white woman was admitted to the hospital after experiencing syncopal episodes secondary to anemia. The patient was status postcholecystectomy, followed by endoscopic retrograde cholangiopancreatography 3 times for retained gallstones. She had episodes of melena and hematochezia after these procedures. The last endoscopic retrograde cholangiopancreatography was performed 1 week prior to this admission. The patient had a history of easy bruising and menorrhagia. Bleeding time was prolonged and lasted 15 minutes, and her platelet count was normal. The peripheral smear demonstrated abnormal platelet morphology with agranular and large forms (Figure 1, arrows). Normal platelets were also present on the blood smear. A platelet aggregation study showed abnormally poor responses to arachidonic acid, adenosine diphosphate, collagen, and epinephrine, and a normal response to ristocetin. Electron microscopy of platelets (Figures 2 and 3) showed 2 populations: one with normal distribution of  $\alpha$  granules and  $\delta$  (dense) granules, the other with marked decrease in both  $\alpha$  granules and  $\delta$  granules (Figure 3). The laboratory findings and clinical history were consistent with storage pool defi-

ciency (SPD), including both  $\alpha$  granule and  $\delta$  granule deficiency. The patient subsequently received platelet and red blood cell transfusions. She was discharged from the hospital 3 days later in stable condition.

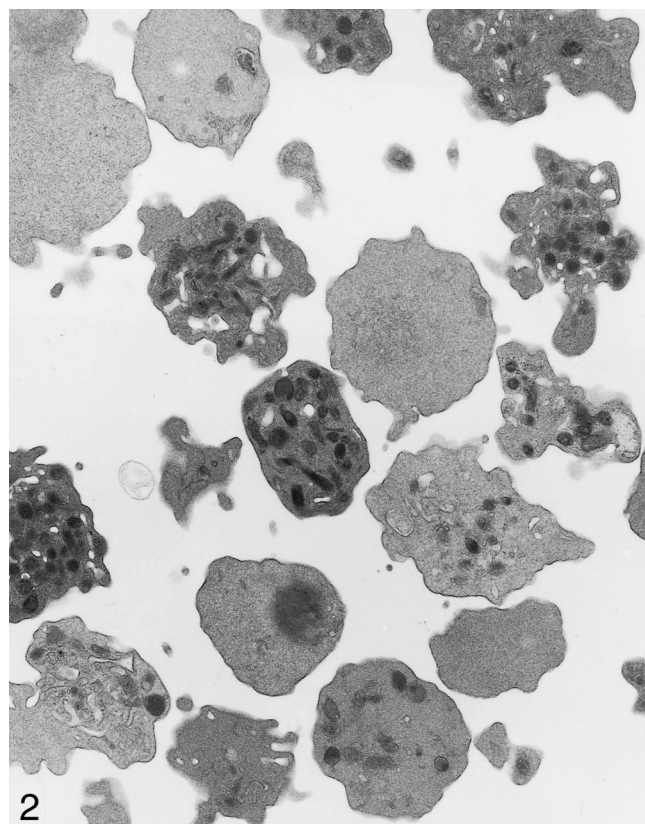
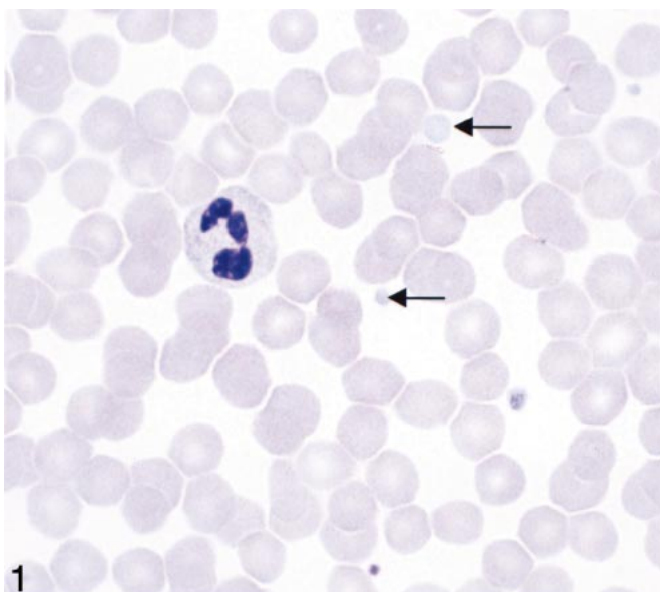
$\alpha$  granules contain a number of different proteins, including fibrinogen, platelet-derived growth factor, von Willebrand factor, factor V, fibronectin,  $\beta$ -thromboglobulin, and heparin-neutralizing factor (platelet factor 4).<sup>1,2</sup>  $\delta$  Granules contain calcium, serotonin, pyrophosphate, adenosine diphosphate, and adenosine triphosphate.<sup>1,2</sup> Determination of storage pool organelles by transmission electron microscopy allows the identification of storage pool defects. On electron microscopy,  $\delta$  granules are electron opaque owing to their high calcium content.<sup>3</sup>

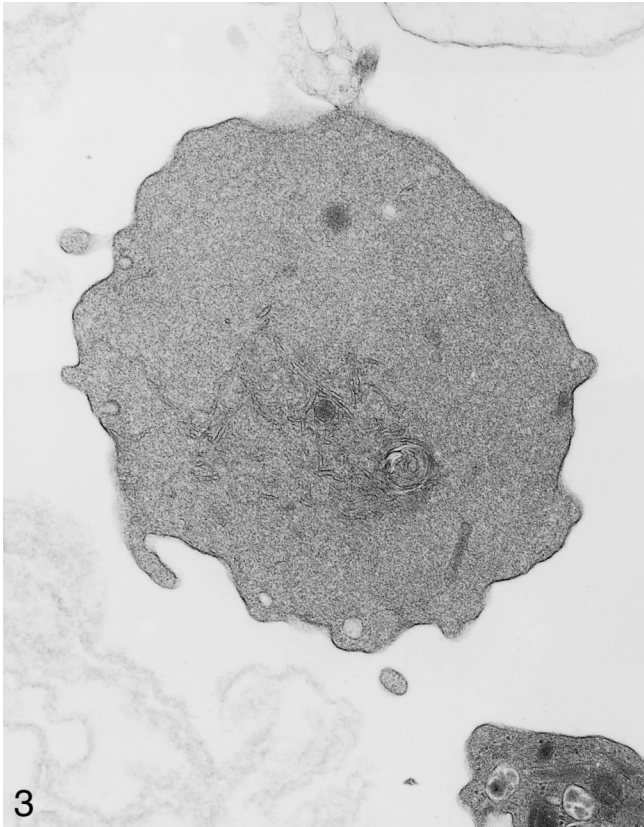
Platelet storage pool deficiencies comprise a range of disorders with variable degrees of reduction in the numbers and contents of  $\alpha$  granules,  $\delta$  granules, or both types of granules.<sup>1,3</sup> The term  $\delta$ -SPD has been used to identify patients who show only a diminished number of  $\delta$  granules. Patients with normal numbers of  $\delta$  granules but decreased

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$\alpha$  granules are designated  $\alpha$ -SPD. Patients with both  $\alpha$ - and  $\delta$ -granule defects are designated  $\alpha\delta$ -SPD. Patients with  $\alpha$ -SPD were originally described as having a "gray platelet syndrome" because of the agranular appearance of their platelets on peripheral blood smears. However, platelets from patients with  $\alpha\delta$ -SPD also appear "gray" on blood smear owing to  $\alpha$ -granule deficiency. Electron microscopic studies have shown that the large vacuoles commonly filling the cytoplasm of  $\alpha$ -SPD platelets are virtually absent from platelets with the combined  $\alpha\delta$  defect.<sup>2</sup>

In most patients with platelet SPD, the platelets aggregate initially to adenosine diphosphate or epinephrine, but second-phase aggregation is frequently markedly diminished. Collagen-induced aggregation is also decreased.<sup>2</sup>

In many patients the storage pool defect is the only abnormality detected. However, the defect has also been observed in patients with other congenital abnormalities, including Hermansky-Pudlak syndrome, Chediak-Higashi syndrome, Wiskott-Aldrich syndrome, and the syndrome of thrombocytopenia with absent radius.<sup>1-3</sup>

#### References

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