Chediak-Higashi syndrome

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An African Kenyan female infant was born with very light skin and ashen grey, scanty hair. At 18 months she presented with a bluish skin pigmentation, hepatosplenomegaly, generalised lymphadenopathy and non-responsive fever. A bone marrow aspirate and peripheral blood examination done revealed characteri¬stic features of the Chediak-Higashi Syndrome. This is a rare disorder, to our knowledge not previously described in Africans. The case is presented with a brief review of the literature