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Waardenburg's syndrome associated with total intestinal aganglionosis

Sir,

Farndon and Bianchi¹ reported a Pakistani child with signs of Waardenburg's syndrome associated with total aganglionosis and suggested that this can be a distinct clinical entity with an autosomal recessive mode of inheritance. We reported our experience of 12 such cases and felt that this was a new syndrome with autosomal recessive inheritance.² Later, we came across another patient who was related to family 5 in our previous report² and this extended pedigree was published.³ On the basis of our

study of this extended pedigree, we have no doubt that inheritance in this syndrome is autosomal recessive while Waardenburg's is an autosomal dominant condition. Farndon's case report lends further support to our impression that this is a distinct clinical entity. It is interesting to note that there are no similar case reports from other parts of the world and we wonder whether any racial factors are involved since the syndrome has been reported only in children of Indian and Pakistani origin to date.

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