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Editorial Note on Bannayan–Riley–Ruvalcaba syndrome (BRRS)

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Editorial

BRRS is an autosomal dominant genodermatosis marked by gastrointestinal hamartomatous polyps, macrocephaly, glans penis hyperpigmentation, developmental delay, and hemangiomas. In 60% of people, a mutation in the PTEN gene can be found in their DNA. This syndrome was first reported as a triad of macrocephaly, lipomatosis, and glans penis coloration. Although BRRS shares certain characteristics with CS, it is typically diagnosed at a younger age, with a male predominance of 68 percent. CS, on the other hand, is more common in women and arises later in life. Hamartomatous polyps, which are seen in 50% of patients, diarrhoea, intussusceptions, and anaemia are all GI symptoms. Polyps can be found throughout the GI tract, however they are more common in the distal ileum and colon.

Although BRRS is not linked to CRC or other GI cancers, persons with PTEN mutations are at risk for cancers of the breast, thyroid, kidney, and endometrial. Hyperpigmented macules involving the glans penis or vulva are the most distinctive cutaneous sign of BRRS. Genital lentigines, facial verrucae-like or acanthosis nigricans-like lesions, multiple acrochordons of the neck, axilla, and groyne, vascular abnormalities, and lipomas are some of the other skin abnormalities. The hyperpigmented lesions are histologically described as lentiginous epidermal hyperplasia, with an increase in the number of melanosomes and a modest rise in melanocytes.

Hypotonia, delayed psychomotor development, seizures, and visual abnormalities involving the retina and cornea are among the other documented findings. Malignancies are a danger for all patients with BRRS, regardless of phenotypic manifestation. As a result, comprehensive management that focuses on early detection through frequent screening of cancer-prone organs is essential. Multiple subcutaneous lipomas, macrocephaly, and hemangiomas are common in Bannayan–Riley–Ruvalcaba syndrome (BRRS), a rare overgrowth syndrome and hamartomatous condition. The disease is passed down through the generations in an autosomal dominant pattern. The disorder is part of a group of hamartomatous polyposis syndromes that includes Peutz–Jeghers syndrome, juvenile polyposis, and Cowden syndrome, among others.

Bannayan–Riley–Ruvalcaba syndrome is linked to enlargement of the skull and benign mesodermal hamartomas. Dysmorphia and delayed neuropsychomotor development are possible side effects. The enlargement of the skull does not result in a widening of the ventricles or an increase in intracranial pressure; nonetheless, because the gene involved in BRRs is phosphatase and tensin homologue, these people are at a higher risk of developing malignancies.

The PTEN gene, which has roughly 30 mutations in this disorder, is used to determine the genetics of the Bannayan–Riley–Ruvalcaba syndrome in the majority of instances. When this gene, which controls cell development, malfunctions, it might cause hamartomas. PTEN is found on chromosome 10q23.31 and on the molecular level at 87,863,438 to 87,971,930. Bannayan–Zonana syndrome, Riley–Smith syndrome, and Ruvalcaba–Myhre–Smith syndrome are all combined in this syndrome. George A. Bannayan and Jonathan Zonana are the names of the Bannayan–Zonana syndrome. There is no existing way for identifying Bannayan–Riley–Ruvalcaba syndrome other than physical traits that may be present as signs/symptoms.

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