

## Opis choroby \*

### Definicja

A rare developmental defect during embryogenesis characterized by hamartomatous intestinal polyposis, lipomas, macrocephaly and genital lentiginosis.

### Dane

|                       |                                   |
|-----------------------|-----------------------------------|
| <b>Klasyfikacja</b>   | <b>Synonimy</b>                   |
| Zespół wad wrodzonych | BRRS                              |
|                       | BRRS                              |
|                       | Zespół de Myhre, Rikey'a i Smitha |
|                       | Zespół Myhre, Riley'a i Smitha    |
|                       | Myhre-Riley-Smith syndrome        |

|                  |                 |                  |
|------------------|-----------------|------------------|
| <b>Kod ORPHA</b> | <b>Kod OMIM</b> | <b>Kod ICD10</b> |
| 109              | 158350          | Q87.8            |

**Kod ICD11**  
LD2D.Y

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### \*Źródło

orphanet