**Table I**. Clinical characteristics and treatment

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| --- | --- | --- | --- |
| **Clinical characteristics** |  |  | **Number of cases** |
| **Number of patients** |  |  | **24** |
| **Gender** | MaleFemale |  | 1212 |
| **Age at diagnosis** | 1-23 years (Median: 13.9) |  > 10 years  ≤ 10 years | 195 |
| **Pathogenic variant** | **Somatic *CTNNB1*** *p.T41A* *p.S45 (F or P)* *Unspecified****APC*** - Germline (FAP) - SomaticNot analysed |  | **17** 10 6 1**4** 3 1**3** |
| **Tumour site** | LimbsTrunkHead - NeckPelvis Abdominal wall |  |  16 (1\*) 4 (2\*)211 |
| **Maximum Tumour size** | ≥ 5 cm< 5 cm |  | 21 3 |
| **Therapies** |  |  |  |
| **Previous therapies** **before oral VNB** | Local therapy: Biopsy Partial resectionRadiotherapySystemic therapy One line Two lines   |  | **24** 20 4 0**20** 18 2  |
| **Wash out period before starting oral VNB** | **0-60 months** (Median: 8.5) | Yes < 12 months ≥ 12 monthsNo | **17** 11 6**7** |
| **Main reason for starting oral VNB** | Tumour progression Radiological Clinical  |  | **24** 21  3  |
| **Schedule of oral VNB** | 60 mg/m²/dose60 mg/m²/dose90 mg/m²/dose 110 mg/m²/dose30-45 mg/m² | **Frequency**4 weeks/month3 weeks/month4 weeks/month3-4 weeks/month 2-3 days/week | 13133 2 |
| **Duration of oral VNB therapy** | **1-36 months** (Median: 12)  |  ≥ 12 months < 12 months | 1311 |
| **Reasons for stopping oral VNB** | End of treatmentTumour progressionPersistent side effects |  | 1725 |