**Table I**. Clinical characteristics and treatment

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| --- | --- | --- | --- |
| **Clinical characteristics** |  |  | **Number of cases** |
| **Number of patients** |  |  | **24** |
| **Gender** | Male  Female |  | 12  12 |
| **Age at diagnosis** | 1-23 years  (Median: 13.9) | > 10 years  ≤ 10 years | 19  5 |
| **Pathogenic variant** | **Somatic *CTNNB1***  *p.T41A*  *p.S45 (F or P)*  *Unspecified*  ***APC***  - Germline (FAP)  - Somatic  Not analysed |  | **17**  10  6  1  **4**  3  1  **3** |
| **Tumour site** | Limbs  Trunk  Head - Neck  Pelvis  Abdominal wall |  | 16 (1\*)  4 (2\*)  2  1  1 |
| **Maximum Tumour size** | ≥ 5 cm  < 5 cm |  | 21  3 |
| **Therapies** |  |  |  |
| **Previous therapies**  **before oral VNB** | Local therapy:  Biopsy  Partial resection  Radiotherapy  Systemic 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 months  No | **17**  11  6  **7** |
| **Main reason for starting oral VNB** | Tumour progression  Radiological  Clinical |  | **24**  21  3 |
| **Schedule of oral VNB** | 60 mg/m²/dose  60 mg/m²/dose  90 mg/m²/dose  110 mg/m²/dose  30-45 mg/m² | **Frequency**  4 weeks/month  3 weeks/month  4 weeks/month  3-4 weeks/month  2-3 days/week | 13  1  3  3  2 |
| **Duration of oral VNB therapy** | **1-36 months**  (Median: 12) | ≥ 12 months  < 12 months | 13  11 |
| **Reasons for stopping oral VNB** | End of treatment  Tumour progression  Persistent side effects |  | 17  2  5 |