hydrocephalus and a non-metastatic lesion in the posterior fossa. A preoperative ECG showed isodifasic T waves in leads V4R and V1, follow-up was recommended. Histology after gross total resection was consistent with Desmoplastic/Nodular Medulloblastoma. At chest X-rays a bifid rib was noted, leading to the diagnosis of Gorlin syndrome (GS; c.3306 + 1G>T in *PTCH1*). During chemotherapy, ventricular tachycardia (VT) occurred, requiring synchronized electrical cardioversion. An echocardiogram revealed an echogenic mass of the left ventricle free wall, but normal coronaries and ventricular function. Cardiac MRI (CMR) confirmed a 21x40x38mm mass, with eccentric development and intense homogeneous enhancement, indicative of cardiac fibroma (CF). A computed tomography excluded local calcifications. Therapy with amiodarone and beta-blocker was initiated. TREATMENT STRATEGY: priority to chemotherapy *vs* cardiac surgery; full-dose chemotherapy, preferring drugs with minor cardiotoxicity; administration in ICU under continuous vital parameters and ECG-monitoring. Other three VTs occurred during treatment or anesthesia, resolved after electrical cardioversion (unsuccessful attempts with i.v. adenosine and amiodarone). Lacking specific guidelines concerning CF in GS, a wait-and-see approach was preferred with close tumor follow-up and regular cardiological assessment (ECG, stress-test, Holter monitoring, CMR). No further arrhythmias were recorded in a 10-year-long follow-up and CMR confirmed CF stability. Medulloblastoma has never recurred. *PTCH1* variants are rarely associated with medulloblastoma (<2%). Only 3–5% of GS present CF, responsible for arrhythmias in 32%. Being non-regressing, total surgical resection is usually performed (without recurrence), with a 27-year-long median survival. When surgery risk/benefit ratio is not favorable or the patient is paucisymptomatic, the treatment plan remains unclear; probably, a conservative approach under a strict cardiological follow-up can be reasonable. In young children with syndromic medulloblastoma, a routine echocardiogram should be performed to rule out CF.

### NFB-12. EFFECT OF TRAMETINIB ON LEG LENGTH DISCREPANCY IN A CHILD WITH NF1 RELATED PLEXIFORM NEUROFIBROMA

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INTRODUCTION: Plexiform neurofibroma(PN) is a challenging benign tumor. Recently, MEK inhibitors (MEKi) showed encouraging tumor response. We report the observed effect of Trametinib on leg length discrepancy (LLD) in a child with NF1. CASE DESCRIPTION: A 4 year old girl with sporadic NF1, developed progressive bilateral L1-L5 paraspinal PN extending to the left thigh resulting in hypertrophy of left leg and associated with LLD. At 33 months of age, length difference of 2.8 cm between both femurs was described on scanogram with a projected LLD of at 6.1- 6.2 cm LLD at bone maturity using the multiplier method, a common method of predicting LLD. At 36 months of age, treatment with Trametinib was initiated for her large PN. Ten months into therapy, parents reported impression of decrease swelling of her left thigh enlargement. MRI evaluation showed stable measurement of PN using the RECIST criteria. Repeat measurement on scanogram at 46 months of age disclosed a stable difference of 2.8 cm between both femurs, with a LLD projected at 5.2-5.3 cm at maturity by multiplier method. Bone age at study entry and at 11 months into therapy (Greulich-Pyle) was reported normal for chronologic age. DISCUSSION/CONCLUSION: LLD has not been commonly described in association with NF1 related PN. Given the PN involved mainly the left thigh in our patient, it is reasonable to suggest common underlying mechanism for the PN and faster growth of her left femur. Although with limited time point's measurements, the early observation of stabilization of the LLD, 10 months into Trametinib therapy suggesting a final discrepancy in femurs length less than initially predicted, is encouraging. Further evaluation at completion of treatment and on follow-up are needed. Larger case series will be useful to explore this unexpected and possible clinical effect of MEKi in NF1 children.

### NFB-13. RHABDOID TUMOR PREDISPOSITION SYNDROME (RTPS) – FINDING EVIDENCE BY SYSTEMATIC ANALYSES

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BACKGROUND: Individuals with rhabdoid tumor predisposition syndrome (RTPS1 – *SMARCB1*, RTPS2 – *SMARCA4*) have a propensity to develop malignant rhabdoid tumors (MRT). Affected patients typically present < age 12 months with synchronous tumors (SYN) exhibiting an unusually aggressive clinical behavior. Due to the rarity of RTPS, standards for management are evolving. METHODS: Clinical, genetic, and treatment data of 90 patients with RTPS from 16 countries were analyzed (2004 – 2020). Therapy followed the EU-RHAB recommendations. Tumors and matching blood samples were investigated for *SMARCB1* and/or *SMARCA4* mutations using FISH, MLPA and sequencing. DNA-methylation subgroups were determined using DNA methylation arrays. RESULTS: The median age at diagnosis of 52 girls and 38 boys was 5.5 months (0 – 203). 55.5% (50/90) of patients