second line treatment- 4 are alive. Children younger than 3 years of age treated without radiotherapy had worse outcome with 5 yrs OS estimated at 0% vs 74,1% in older children treated with chemotherapy and radiotherapy (p=0,014). CONCLUSION: Pineoblastoma is highly malignant CNS neoplasm of childhood with dismal prognosis. In our series children below 3 years of age treated without radiotherapy had poorer responses and worse outcome.

CRAN-07. SIGNIFICANT RATES OF ACTH SUPPRESSION MISDIAGNOSED AS ACTH DEFICIENCY AFTER PERIOPERATIVE DEXAMETHASONE IN CRANIOPHARYNGIOMA

Kyriaki Pieri, Maria Michaelidou, Antonia Dastamani, and Helen A Spoudeas; London Centre for Paediatric Endocrinology, Great Ormond Street (GOSH) and University College Hospitals (UCLH), London, UK

INTRODUCTION: ACTH suppression may masquerade as ACTH deficiency in children and young people (CYP) receiving dexamethasone for pituitary tumour surgery. In our experience, GH is always the first, and ACTH the last anterior pituitary deficit, with LH/FSH and TSH intermediate in hierarchical loss. AIMS: To investigate long term adrenal recovery and true ACTH deficiency rate in patients with craniopharyngioma receiving hydrocortisone replacement, and whether a spontaneous puberty and detectable pre-dose 8am plasma ACTH predict intact ACTH reserve. METHODS: Longitudinal retrospective record review of 53 cases (29M), diagnosed aged 6.87(1.12–17.18) years and followed for 6.98(1.43–26.73) years. RESULTS: 9 of 53 (16.98%) never required hydrocortisone of whom 8 (88.9%) were GHD, 6 (66.7%) TSHD, 5 (55.6%) LH/FSHD and 1 (11.1%) had DI at last follow-up. A further 6 of 53 (11.3%) had hydrocortisone for presumed ACTHD (peak cortisol <117nmol/l) but "recovered" with intact cortisol reserve and detectable plasma ACTH 36.2ng/L(13.9–52.9) after 3.08(2.38–10.33) years. All were GHD, 5 (83.3%) LH/FSHD, 4 (66.7%) TSHD and 2 (33.3%) had DI. In the last 38 (71.7%) patients, ACTH remains undetectable (<5ng/L); all require cortisol, GH and thyroxine replacement, 31/32 (96.9%) of postpubertal age require sex steroids and 31/38 have DI, after 6.40(1.43–26.73) years. CONCLUSIONS: Adrenal suppression can be prolonged. Significant (13.6%) recovery rates at a median 7 years may further increase with time. Overdiagnosis of ACTHD may be misattributed to surgery or radiation. We encourage readdressing the need for hydrocortisone in CYP with detectable pre-dose 8am ACTH >10ng/L and without concurrent TSH and post-pubertal LH/FSH deficiency.

CRAN-08. NATIONAL UK GUIDELINES FOR THE INVESTIGATION, TREATMENT AND LONG-TERM FOLLOW-UP OF PAEDIATRIC CRANIOPHARYNGIOMA

Hoong-Wei Gan[1,3], Kristian Aquilina[3], Paul Morillon[2], Assunta Albanese[8], Konstantinos Barkas[9], Chris Chandler[9], Yen-Ching Chang[10], Christina Daousi[11], Evangelos Drimtzias[12], Sarah Farndon[13], Tom Jacques[3], Marta Korbonits[14], Adam Kuczynski[3], Jennifer Limond[15], Louise Robinson[16], Ian Simmons[12], Nick Thomas[9], Sophie Thomas[6], Nicola Thorpe[17], Faraneh Vargha-Khadem[1,3], Daniel Warren[12], Bassel Zebian[9], Conor Mallucci[4], Ashley Gamble[5], Sophie Wilne[6], Barney Harrison[7], and Helen Spoudeas[3]; [1]University College London Great Ormond Street Institute of Child Health, London, UK, [2]Kingston Hospital NHS Foundation Trust, London, UK, [3]Great Ormond Street Hospital for Children NHS Foundation Trust, London, UK, [4]Alder Hey Children's NHS Foundation Trust, Liverpool, UK, [5]UK Children's Cancer and Leukaemia Group, Leicester, UK, [6]Nottingham Children's Hospital, Nottingham University Hospitals NHS Trust, Nottingham, UK, [7]Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK, [8]St. George's University Hospitals NHS Foundation Trust, London, UK, [9]King's College Hospital NHS Foundation Trust, London, UK, [10]University College London Hospitals NHS Foundation Trust, London, UK, [11]Aintree University Hospital NHS Foundation Trust, Liverpool, UK, [12]Leeds Teaching Hospitals NHS Trust, Leeds, UK, [13]East and North Hertfordshire NHS Trust, Stevenage, UK, [14]Queen Mary University of London, London, UK, [15]University of Glasgow, Glasgow, UK, [16]Royal Manchester Children's Hospital, Central Manchester University Hospitals NHS Foundation Trust, Manchester, UK, [17]The Clatterbridge Cancer Centre NHS Foundation Trust, Birkenhead, UK

BACKGROUND: Craniopharyngiomas are the commonest suprasellar tumour in childhood. Despite high overall survival, children with craniopharyngiomas are at risk of multiple relapses and long-term tumour- and treatment-related morbidity. We sought to provide, for the first time, a national evidence-based standard for best practice for the assessment, treatment, and follow-up of paediatric craniopharyngiomas. METHODS: 44 clinical questions were formulated based on a PICO (Population, Intervention, Comparison, Outcome) format by a multidisciplinary Guideline Development Group to guide systematic searches via the Ovid MEDLINE (1946-February 2017) and Cochrane Library (2016, Issue 12) databases, identifying 2023 separate research articles. Publications underwent a three-tier filtering process and 300 were reviewed using the GRADE approach. Where recommendations could not be made, a two-stage international Delphi consensus process was conducted. RESULTS: 35 recommendations were made, largely based on low to very low quality evidence. 30 further recommendations achieved >70% agreement via Delphi consensus. Craniopharyngiomas should be managed in tertiary paediatric centres with sufficient neuro-oncology, neurosurgery, endocrinology, radiology, pathology and neuropsychology experience. At diagnosis, tumours should be graded using the "Paris" grading system and surgical treatment tailored to avoid hypothalamic damage, with adjuvant radiotherapy being offered after incomplete resections. Recommendations on the long-term multidisciplinary follow-up of survivors are also detailed. CONCLUSIONS: These Royal College of Paediatrics and Child Health (RCPCH)-endorsed guidelines provide the first evidence-based national recommendations for the management of paediatric craniopharyngioma. Through their implementation, we hope to achieve better consistency in the quality of care of such patients and improve long-term quality of survival.

CRAN-09. CHOROID PLEXUS CARCINOMA IN INFANCY: FAVORABLE OUTCOME IN THREE PATIENTS TREATED WITH CHEMOTHERAPY

Reuben Antony[1], James Boggan[1], Arzu Ozturk[1], Theo Nicolaides[2], Lee-Way Jin[1], and Marike Zwienenberg-Lee[1]; [1]University of California at Davis Comprehensive Cancer Center, Sacramento, CA, USA, [2]University of California, San Francisco, San Francisco, CA, USA

BACKGROUND: Children diagnosed with Choroid Plexus Carcinoma (CPC) in the first year of life usually have a very poor outcome partly because radiotherapy cannot be used as a part of treatment. We report 3 infants diagnosed with CPC in the first year of life including one patient who has Li-Fraumeni Syndrome who were successfully treated solely with chemotherapy. CASE REPORTS: Patient 1: diagnosed at age 1.5 months with a non-metastatic CPC in the 3rd ventricle. Following gross total tumor resection (GTR) he was treated on COG protocol POG 9923/34 using Vincristine, Etoposide, Cyclophosphamide and Cisplatin (VECC). He remains disease free 4 years post completion of therapy. Patient 2: diagnosed at age 8 months with a non-metastatic left lateral ventricle CPC. Following GTR he was treated on protocol POG 9932/34. He remains disease free 6 years post completion of therapy. Patient 3: Patient had a strong family history of cancer and was diagnosed with an adrenocortical neoplasm at birth. She was diagnosed at age 6 months with a right lateral ventricle CPC disseminated through the brainstem and spinal cord. After a GTR of the intracranial tumor she was treated as per the Head Start 2 (Regimen A) protocol using VECC and High Dose Methotrexate followed by triple autologous stem cell transplant (Carboplatin/Thiotepa conditioning). She remains disease free 8 months post completion of therapy. All patients experienced significant bacterial sepsis episodes, malnutrition and ototoxicity. CONCLUSION: Long term tumor control of CPC is possible using chemotherapy only protocols in very young infants.

CRAN-10. PEDIATRIC CRANIOPHARYNGIOMA IN ASSOCIATION WITH FAMILIAL ADENOMATOUS POLYPOSIS

Nathan A. Dahl[1,2], Drew Pratt[3], Chandan Kumar[4], Sandra Camelo-Piragua[3], Rajen J. Mody[5], Seth Septer[6], Todd Hankinson[2,7], Carl Koschmann[5], and Lindsey Hoffman[1]; [1]Pediatric Neuro-Oncology Program, Children's Hospital Colorado, University of Colorado School of Medicine, Denver, CO, USA, [2]Morgan Adams Foundation Pediatric Brain Tumor Research Program, Denver, CO, USA, [3]Neuropathology Division, Department of Pathology, University of Michigan, Ann Arbor, MI, USA, [4]Center for Translational Pathology, Department of Pathology, University of Michigan, Ann Arbor, MI, USA, [5]Pediatric Hematology-Oncology, Department of Pediatrics, University of Michigan, Ann Arbor, MI, USA, [6]Pediatric Gastroenterology, Children's Hospital Colorado, University of Colorado School of Medicine, Denver, CO, USA, [7]Department of Neurosurgery, Children's Hospital Colorado, University of Colorado School of Medicine, Denver, CO, USA

Familial adenomatous polyposis (FAP) is a cancer predisposition syndrome that is driven by germline loss-of-function of the APC gene and phenotypically characterized by intestinal polyposis and a variety of extra-intestinal bone and soft tissue tumors. Though craniopharyngioma is not classically associated with FAP, six cases have been reported in adults, all demonstrating ectopic location and adamantinomatous histology. We report the first case of craniopharyngioma associated with FAP in a pediatric patient. A seven-year-old girl with known FAP presented with headache and vomiting and was found on head CT to have partially calcified, cystic mass involving the posterior fossa. MRI delineated a primary suprasellar mass with cystic extension to the pre-pontine space. The cyst was surgically decompressed, and pathology demonstrated an adamantinomatous craniopharyngioma with nuclear β-catenin expression. We performed whole exome sequencing of paired germline and tumor DNA, confirming a CTNNB1 activating point