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High prevalence of adrenal insufficiency at diagnosis and headache recovery in surgically resected Rathke's cleft cysts-a large retrospective single center study.

Langlois F(1), Manea A(2), Lim DST(3), McCartney S(4), Yedinak CG(4), Cetas JS(4), Fleseriu M(5)(6).

Author information:

(1)Department of Endocrinology, Centre hospitalier universitaire de Sherbrooke, Fleurimont, QC, Canada.

(2)Pediatric Endocrinology, Oregon Health & Science University, Portland, OR, USA.

(3)Department of Endocrinology, Singapore General Hospital, Singapore, Singapore.

(4)Neurological Surgery, Oregon Health & Science University, Portland, OR, USA.

(5)Neurological Surgery, Oregon Health & Science University, Portland, OR, USA. fleseriu@ohsu.edu.

(6)Pituitary Center, Medicine and Neurological Surgery, Oregon Health & Science University, Portland, OR, USA. fleseriu@ohsu.edu.

BACKGROUND: Rathke's cleft cysts (RCC) are lesions that arise from Rathke's pouch. Though frequently incidental, resulting symptoms in a minority of cases are indicators for surgical resection, which may prove beneficial.

OBJECTIVE: To characterize a cohort of surgically-resected RCC cases at Oregon Health & Science University; tabulate associated hormonal imbalances and symptoms, possible symptom reversal with surgery, determine recurrence risk; identify predictors of recurrence and headache improvement.

METHOD: Electronic records of all RCC resected cases (from 2006-2016; 11 years) were retrospectively reviewed. Patients had been evaluated by one neuroendocrinologist using a uniform protocol.

RESULTS: A pathological RCC diagnosis was established in 73 of 814 (9%) surgical pituitary cases. The RCC cohort was 77% (n = 56/73) female, mean age was 39.5 ± 14.9 years at first surgery, and at presentation headache was reported in 88% and visual defects/diplopia in 18% of patients. Initial RCC maximum diameter was 1.3 ± 0.7 cm. The most frequent hormonal deficit was cortisol; 24% of patients had a new adrenal insufficiency (AI) diagnosis, however, 36% also had AI at 3 months post-operatively. Mean follow up was 4.0 ± 4.5 years. Two-thirds of patients (41/62) had headache improvement 3 months post-operatively.

Post-operative imaging revealed no residual cyst in 58% (38/65). In those patients with no residual RCC, 29% had recurrence and 71% had long lasting cure. From the 42% (27/65) of patients with residual cyst on post-operative imaging; 59% (16/27) remained stable, 26% (7/27) progressed and 15% (4/27) regressed.

CONCLUSION: Symptomatic RCC present mostly in women, with a high proportion reporting headaches. Prevalence of AI at diagnosis is high. Surgery may not achieve adrenal axis recovery, but renders a high percentage of headache improvement. Approximately 25% of RCC will recur by 4 years postoperatively. Clinicians should cautiously screen patients with symptomatic RCC, regardless of lesion size for AI.

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Radiation exposure of adrenal vein sampling: a German Multicenter Study

Fuss CT(1), Treitl M(2), Rayes N(3), Podrabsky P(4), Fenske WK(5)(6), Heinrich DA(7), Reincke M(7), Petersen TO(8), Fassnach M(1), Quinkler M(9), Kickuth R(10), Hahner S(1).

Author information:

(1)Department of Medicine I, Division of Endocrinology and Diabetology, University Hospital, University of Würzburg, Würzburg, Germany

(2)Department of Clinical Radiology, Ludwig-Maximilians-University, Munich, Germany

(3)Department of General, Visceral and Transplant Surgery, University Hospital Leipzig, Leipzig, Germany

(4)Department of Radiology, Charité Campus Virchow Klinikum, Berlin, Germany

(5)Leipzig University Medical Center, Integrated Center for Research and Treatment Adiposity Diseases, Leipzig, Germany

(6)Internal Medicine (Endocrinology and Nephrology), University of Leipzig, Leipzig, Germany

(7)Department of Endocrinology, Ludwig-Maximilians-University, Munich, Germany

(8)Department of Diagnostic and Interventional Radiology, University Hospital Leipzig, Leipzig, Germany

(9)Endokrinologie in Charlottenburg, Endokrinologie Praxis am Stuttgarter Platz, Berlin, Germany

(10)Department of Radiology, University Hospital Würzburg, Würzburg, Germany

Objective: Adrenal vein sampling (AVS) represents the current diagnostic standard for subtype differentiation in primary aldosteronism (PA). However, AVS has its drawbacks. It is invasive, expensive, requires an experienced interventional radiologist and comes with radiation exposure. However, exact radiation exposure of patients undergoing AVS has never been examined.

Design and Methods: We retrospectively analyzed radiation exposure of 656 AVS performed between 1999 and 2017 at four university hospitals. The primary outcomes were dose area product (DAP) and fluoroscopy time (FT). Consecutively the effective dose (ED) was approximately calculated.

Results: Median DAP was found to be 32.5 Gy*cm² (0.3–3181) and FT 18 min (0.3–184). The calculated ED was 6.4 mSv (0.1–636). Remarkably, values between participating centers highly varied: Median DAP ranged from 16 to 147 Gy*cm², FT from 16 to 27 min, and ED from 3.2 to 29 mSv. As main reason for this variation, differences regarding AVS protocols between centers could be identified, such as number of sampling locations, frames per second and the use of digital subtraction angiographies.

Conclusions: This first systematic assessment of radiation exposure in AVS not only shows fairly high values for patients, but also states notable differences among the centers. Thus, we not only recommend taking into account the risk of radiation exposure, when referring patients to undergo AVS, but also to establish improved standard operating procedures to prevent unnecessary radiation exposure.

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Genital Reconstructive Surgery in Females With Congenital Adrenal Hyperplasia: A Systematic Review and Meta-Analysis.

Almasri J(1)(2), Zaiem F(3), Rodriguez-Gutierrez R(4)(5), Tamhane SU(6), Iqbal AM(7), Prokop LJ(8), Speiser PW(9), Baskin LS(10), Bancos I(6), Murad MH(1)(2).

Author information:

(1)Evidence-Based Practice Research Program, Mayo Clinic, Rochester, Minnesota.

(2)Robert D. and Patricia E. Kern Center for the Science of Health Care Delivery, Mayo Clinic, Rochester, Minnesota.

(3)Department of Pathology, Karmanos Cancer Institute, Wayne State University School of Medicine, Detroit, Michigan.

(4)Division of Endocrinology, Department of Internal Medicine, University Hospital Dr. Jose E. Gonzalez, Autonomous University of Nuevo Leon, Monterrey, Mexico.

(5)Plataforma INVEST Medicina UANL-KER Unit Mayo Clinic, Autonomous University of Nuevo Leon, Monterrey, Mexico.

(6)Division of Endocrinology, Mayo Clinic, Rochester, Minnesota.

(7)Division of Pediatrics and Adolescent Medicine, Department of Pediatric Endocrinology, Mayo Clinic, Rochester, Minnesota.

(8)Mayo Clinic Libraries, Mayo Clinic, Rochester, Minnesota.

(9)Division of Pediatric Endocrinology, Cohen Children's Medical Center and Zucker Hofstra Northwell School of Medicine, Lake Success, New York.

(10)Department of Urology, University of California, San Francisco, California.

Background: Females with congenital adrenal hyperplasia (CAH) and atypical genitalia often undergo complex surgeries; however, their outcomes remain largely uncertain.

Methods: We searched several databases through 8 March 2016 for studies evaluating genital reconstructive surgery in females with CAH. Reviewers working independently and in duplicate selected and appraised the studies.

Results: We included 29 observational studies (1178 patients, mean age at surgery, 2.7 ± 4.7 years; mostly classic CAH). After an average follow-up of 10.3 years, most patients who had undergone surgery had a female gender identity (88.7%) and were heterosexual (76.2%). Females who underwent surgery reported a sexual function score of 25.13 using the Female Sexual Function Index (maximum score, 36). Many patients continued to complain of substantial impairment of sensitivity in the clitoris, vaginal penetration difficulties, and low intercourse frequency. Most patients were sexually active, although only 48% reported comfortable intercourse. Most patients (79.4%) and treating health care professionals (71.8%) were satisfied with the surgical outcomes. Vaginal stenosis was common (27%), and other surgical complications, such as fistulas, urinary incontinence, and urinary tract infections, were less common. Data on quality of life were sparse and inconclusive.

Conclusion: The long-term follow-up of females with CAH who had undergone urogenital reconstructive surgery shows variable sexual function. Most patients were sexually active and satisfied with the surgical outcomes; however, some patients still complained of impairment in sexual experience and satisfaction. The certainty in the available evidence is very low.

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Robotic Adrenalectomy: Are We Expanding the Indications of Minimally Invasive Surgery?

Quadri P(1), Esposito S(2), Coleoglou A(1), Danielson KK(1)(2), Masrur M(1), Giulianotti PC(1).

Author information:

(1)1 Division of General, Minimally Invasive and Robotic Surgery, Department of Surgery, University of Illinois at Chicago , Chicago, Illinois.

(2)2 Division of Epidemiology and Biostatistics, University of Illinois at Chicago , Chicago, Illinois.

INTRODUCTION: Laparoscopic adrenalectomy (LA) is accepted as the gold standard treatment for most adrenal pathologies. Open surgery is still considered the standard of care for large tumors and malignancies. In the past decade, robotic adrenalectomy (RA) has become an alternative to the laparoscopic and open approaches. The aim of this study was to analyze perioperative and postoperative outcomes in a series of consecutive nonselected patients undergoing a RA, to determine whether factors that negatively affect outcomes in LA (body mass index [BMI], size, and side of the tumor) have the same impact in RA.

MATERIALS AND METHODS: This is a single-center single-surgeon retrospective study with 43 patients who underwent a RA. Patients were divided into different groups according to tumor size (cutoff values of 5 or 8 cm), tumor side (left/right), and BMI (cutoff value of kg/m²). Perioperative and postoperative outcomes included operative time, length of hospital stay, blood loss, readmissions, complications, and conversions to open.

RESULTS: There were no significant differences between the groups with tumors <5 cm versus ≥5 cm regarding gender, age, race, BMI, American Society of Anesthesiologists (ASA) score, history of previous abdominal surgery, tumor side, and histopathological diagnosis (all P values ≥.06). There were no significant differences in any of the outcomes analyzed with respect to the tumor size (all P values ≥.14) except for a higher occurrence of complications in patients with tumors ≥8 cm versus <8 cm (P = .03). There were no significant differences in any outcomes related to side (left versus right) of the tumor nor BMI (<30 versus ≥30 kg/m²). The overall readmission and conversion rates were both 2.3% and no mortalities were registered.

CONCLUSION: Patient's BMI, tumor side, and size did not demonstrate a negative impact on perioperative and postoperative outcomes of RA. This approach could potentially expand the indications of minimally invasive surgery.

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Adrenal Suppression from Topical and Subconjunctival Steroids after Pediatric Cataract Surgery.

Lambert SR(1), Shah S(2).

Author information:

(1)Palo Alto, California. Electronic address: lambert7@stanford.edu.

(2)Palo Alto, California.

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Unusual Techniques for Preserving Surgical and Oncologic Safety in Hepatectomy of Advanced Adrenal Malignancy with Vena Cava and Liver Invasion.

Lee JM(1), Lee KW(2), Hong SK(1), Yoon KC(1), Cho JH(1), Yi NJ(1), Suh KS(1).

Author information:

(1)Department of Surgery, Seoul National University College of Medicine, Seoul, Korea.

(2)Department of Surgery, Seoul National University College of Medicine, Seoul, Korea. kwleegs@gmail.com.

BACKGROUND: Status in terms of major vascular structure invasion is a crucial factor for successful major hepatic resection. In particular, surgery for advanced tumors with inferior vena cava (IVC) invasion is difficult and may even be dangerous for the patient, having high risk of massive bleeding and greater chance of embolic complications such as stroke, bowel ischemia, and pulmonary venous thrombosis. For such reasons, many surgeons hesitate to carry out such surgical resection, and even if they do so, may not totally remove the tumor including the part inside the IVC, achieving R1 resection. For safe and radical surgery, various surgical techniques are required. We report herein three cases of major hepatectomy with IVC invasion and discuss several surgical tips.

PATIENTS AND METHODS: From March 2011 to February 2014, we retrospectively reviewed three cases of adrenal malignancy with liver and IVC invasion. Based on the severity of the malignant tumor, each case illustrates a different method to address surgical complications and maintain oncologic safety. Case 1: A 34-year-old woman was diagnosed with adrenocortical tumor during medical examination. Tumor invaded the right lobe of the liver and very close to the IVC. Fortunately, there was little thrombosis inside the IVC; we performed right hemihepatectomy and adrenalectomy, then resected the IVC wall close to the tumor and repaired the IVC side wall using 4-0 Prolene. Case 2: A 54-year-old woman who complained of abdominal discomfort visited our hospital. Abdominal computed tomography (CT) scan revealed huge adrenal mass with liver and IVC invasion. Thrombosis inside the IVC extended to the right atrium. We decided to carry out veno-veno bypass during operation in collaboration with heart surgeon. After application of veno-veno bypass, the right atrium wall was opened and the tumor thrombus removed. We then carried out right hemihepatectomy and adrenalectomy. Supra- and infrahepatic vena cava were clamped during tumor thrombectomy to prevent embolic complications. Case 3: A 51-year-old woman who complained of headache and hypertension visited our hospital and was diagnosed with huge adrenal tumor. Tumor invaded to the right lobe of the liver and encased the IVC. The tumor totally invaded the IVC, and massive bleeding was expected during dissection. We resected the tumor including IVC en bloc, and reconstructed IVC with artificial graft (Dacron) under veno-veno bypass.

RESULTS: In case 1, there was no surgical complication. The patient was discharged 7 days postoperatively and underwent adjuvant chemotherapy (Mitotane) after discharge. Unfortunately, multiple hepatic metastases were identified 4 months after operation. She died 6 months after surgery. In case 2, there was no

surgical complication after surgery. The patient was discharged 10 days postoperatively. Multiple liver and lung metastases were identified 4 months after operation, and pulmonary embolism was also diagnosed on chemotherapy. She died 16 months after operation. In case 3, the patient had no surgical complication in the immediate postoperative period and was discharged 14 days after surgery. Pheochromocytoma was confirmed in pathologic report. One month after discharge, she underwent interventional balloon dilatation due to short segmental collapse of suprahepatic IVC. At 42 months after surgery, she was still alive with no relapse.

DISCUSSION: In advanced-stage malignant tumor, the conflict between achieving oncologic R0 resection and patient safety remains an unsolved issue. In particular, more advanced surgical technique is required when the tumor invades large vessels such as the vena cava. Previous reports on cases of advanced tumor invading liver and IVC have described the technical difficulties.^{1,2} Wakayama et al. reported cases of successful thrombectomy under veno-veno bypass in hepatocellular carcinoma with IVC and right atrium invasion,³ and Vicente et al.⁴ reported surgical resection of IVC thrombus without cardiopulmonary bypass. Major vascular invasion of the tumor is known to be a poor prognostic factor for survival. However, some reports state that, if the tumor invades major vascular structure, complete tumor removal might be helpful for patient survival due to the biologic features of the tumor.^{2,5,6} This video report does not describe any new techniques, but is more helpful for junior surgeons in educational terms. The limitation of this report is that we could not show good oncologic long-term survival after surgery. However, no fatal complications related to the surgical procedure occurred, by managing the tumor thrombus during the operation. We present three techniques with differing aggressiveness. The techniques illustrated in this video represent a good option to achieve patient surgical safety.

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Arterial stiffness evaluated by pulse wave velocity is not predictive of the improvement in hypertension after adrenal surgery for primary aldosteronism: A multicentre study from the French European Society of Hypertension Excellence Centres.

Bouhanick B(1), Amar J(2), Amar L(3), Gosse P(4), Girerd X(5), Reznik Y(6), Mounier-Vehier C(7), Baguet JP(8), Boutouyrie P(9), Lepage B(10), Lantelme P(11), Chamontin B(12); investigators of the ASAPAS study.

Author information:

(1)Department of Therapeutics and Hypertension, Rangueil University Hospital, 31059 Toulouse, France; UMR 1027 INSERM Toulouse 3 University, 31000 Toulouse, France. Electronic address: duly-bouhanick.b@chu-toulouse.fr.

(2)Department of Therapeutics and Hypertension, Rangueil University Hospital, 31059 Toulouse, France; INSERM 1048 Toulouse 3 University, 31432 Toulouse, France.

(3)Hypertension Unit, hôpital Européen-Georges-Pompidou, université Paris Descartes, Assistance publique-Hôpitaux de Paris, 75908 Paris, France.

(4)Department of Cardiology and Hypertension, Bordeaux University Hospital, 33076

Bordeaux, France.

(5)Unité de prévention cardiovasculaire, University Hospital Pitié-Salpêtrière, 75651 Paris, France.

(6)Department of Endocrinology, Côte de Nacre Regional Hospital Center, University of Caen, 14033 Caen, France.

(7)Service de médecine vasculaire et HTA, University Hospital Lille, 59037 Lille, France.

(8)Department of Cardiology, Mutualist hospital Group, 38100 Grenoble, France.

(9)Inserm, U970, Pharmacology Unit, hôpital Européen Georges-Pompidou, AP-HP, 75908 Paris, France; Cardiovascular Research Center PARCC, University Paris Descartes, Sorbonne Paris Cité, 75015 Paris, France.

(10)UMR 1027 INSERM Toulouse 3 University, 31000 Toulouse, France; Department of Epidemiology, Toulouse University Hospital, 31000 Toulouse, France.

(11)Cardiology Department, hospices civils de Lyon, hôpital de la Croix-Rousse, 69004 Lyon, France.

(12)Department of Therapeutics and Hypertension, Rangueil University Hospital, 31059 Toulouse, France.

BACKGROUND: Predictive factors associated with normal blood pressure (BP) after unilateral adrenalectomy for primary aldosteronism (PA) are not clearly identified.

AIMS: To evaluate the predictive value of arterial stiffness before surgery on BP after surgery.

METHODS: During 2009-2013, 96 patients with PA due to unilateral adrenal adenoma who underwent surgery were enrolled in a multicentre open-label, prospective study. Aortic pulse wave velocity (PWV) was assessed before surgery. Patients underwent ambulatory blood pressure monitoring (ABPM) before surgery and 6 and 12 months after surgery. Twenty-four h SBP/DBP values were compared in subjects with PWV < vs. ≥ 10 m/s. The primary outcome was 24-hour ABPM < 130/80 mmHg 6 months after adrenalectomy.

RESULTS: BP and PWV were available for 82 patients (mean age 49 ± 12 years). Mean 24-hour systolic/diastolic BP (SBP/DBP) values decreased from $144 \pm 15/91 \pm 9$ before surgery to $131 \pm 15/84 \pm 11$ mmHg 6 months after surgery. At 6 months, mean 24-hour SBP did not differ significantly between high versus low PWV groups (SBP -0.8 mmHg, 95% confidence interval -6.9 to 5.2, $P=0.79$). A total of 42.3% of women versus 20.0% of men had 24-hour SBP/DBP < 130/80 mmHg at 6 months ($P=0.07$) and 57.9% vs. 23.8% at 12 months ($P=0.03$). Higher SBP/DBP was recorded for men versus women after 6 months ($P=0.01/0.001$) and 1 year ($P=0.04/0.05$).

CONCLUSION: Preoperative arterial stiffness does not predict a beneficial effect of adrenalectomy on BP values.

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