Testicular masses in association with congenital adrenal hyperplasia:
MR features compared with sonographic findings

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Abstract
Purpose: imaging findings of testicular tumors in patients with congenital adrenal hyperplasia (CAH). Methods and materials: 17 patients with CAH due to 21-OH deficiency were evaluated. All patients underwent ultrasound (US) and in 16 patients MR was performed. T2W and T1W images were obtained. Results: US revealed lesions in 94%. The size varied from 2 mm to 40x20-25 mm. All lesions were hypoechoic and located adjacent to the mediastinum testis. Acoustic shadowing and well-defined margins was present in 16 cases. MR revealed lesions in 94%. On T2W images the lesions appeared as hypo-intense, well defined, homogeneous nodular masses, located adjacent to the mediastinum. All lesions demonstrated strong enhancement after administration of iv gadolinium (Gd). Conclusion: Testicular masses are common in CAH and have characteristic US and MR appearance.

Introduction
In patients with CAH due to 21-OH deficiency, the synthesis of cortisol, and in most cases also of aldosterone, is impaired. Consequently, the secretion of ACTH by the pituitary gland is increased, resulting in hyperplasia of the adrenal cortex and production of high amounts of adrenal androgens. Treatment of CAH consists of substitution of cortisol and aldosterone, thereby suppressing adrenal androgen overproduction [1,2]. Testicular tumors in CAH are most often associated with poorly controlled disease. The prevalence of adrenal testicular adrenal rest tumors in well-controlled patients with CAH is insufficiently known. We investigated the prevalence of testicular tumors in patients with 21-OH deficiency and compared US and MR features of the testicular lesions.

Patients and Methods
17 patients with CAH due to 21-OH deficiency participated in this study. The age of the patients was 21.8 ± 6.5 years (mean ± SD; range 16.6 to 40.8 years). In all patients 21-OH deficiency had been confirmed by DNA-analysis. Imaging of the testes was performed by scrotal ultrasound (US), color flow Doppler and MR imaging. The time interval between ultrasound and MRI varied from 3.7 to 10.4 months (median 5 months). Scrotal US was performed using a 6000 Powervision scanner (Toshiba Medical Systems®, Ottawa, Japan) 7.5 MHz transducer. The testes were evaluated for size, location and echogenicity. Testicular volume was calculated using the formula: V=LxWxDx0.52, where V is the testis volume (ml), L the maximal testicular length (cm), W the maximal width (cm) and D the maximal depth (cm). The MR studies were performed in 16 of the 17 patients using a 1.5-T scanner (Magnetom Vision, Siemens, Erlangen, Germany), and a body-phase-array coil. In all 16 patients, we obtained unenhanced T1W images (TR/TE, 800/12; matrix 150x256; number of excitations, 2), and T2W images (TR/TE, 4400/132; matrix 150x256; number of excitations, 3) of the testes in the saggital plane, with and without fat suppression. In all patients enhanced T1W images were obtained after iv administration of 15 ml Gd (dose: 0.1 mmol/kg over 6 seconds with a flow rate of 2.5 ml/sec). These images were subtracted from the unenhanced.

Results
Testicular volumes ranged from 4.2-26.7 ml (mean ± SD; 11.7 ± 5.2 ml). US depicted testicular lesions in 16 of 17 patients (26 of the 34 testes). In only 6 of the 16 patients the abnormalities were clinically palpable. The tumor size varied from 2 mm to 40x20-25 mm. In 10 patients the lesions were located bilaterally, and in 6 unilaterally. In 9 patients the masses were hypoechoic. The majority of these lesions was smaller than 1 cm (82%). In 7 patients the masses were hypoechoic with hyperechoic reflection. The majority of these lesions was larger than 2 cm (79%). The nodules were mostly multifocal. Hypervascularity was seen in 4 tumors. The other tumors were normovascular. All lesions were located intratesticular, and were found at or around the mediastinum.

MR depicted lesions in 15 of 16 patients. All masses were isointense on T1W, and all masses were hypointense on T2W. None of the masses displayed any evidence of a capsule or pseudocapsule. Following iv of Gd all lesions demonstrated strong enhancement (Fig. 1).

Discussion and Conclusion
Testicular adrenal rest tissue is the main cause of testicular masses in patients with untreated or poorly controlled CAH. In our series of the 17 patients with well controlled CAH had testicular masses detected by radiologic examinations. US typically reveals often bilateral testicular masses located in the region of the mediastinum testis [3]. Testicular tumors generally develop bilaterally in patients with CAH and poor adrenal steroid genesis suppression. Exogenous steroid therapy causes neoplasm regression in most patients with CAH, whereas Leydig cell tumors are unresponsive to steroid. When CAH is undiagnosed and a patient presents with a testicular neoplasm, it is clinically difficult to distinguish testicular tumors of CAH from Leydig cell tumors. The similar histological, morphological and radiographic appearance of these lesions makes differentiation difficult. Neither US nor MR imaging findings are specific for testicular adrenal rest tissue, because some testicular malignancies may have similar features. Once CAH is suspected the patient should be followed up with imaging studies to assess changes in size. Both MR and US detect and display masses composed of testicular adrenal rest tissue equally well [4,5,6]. Testicular masses are common in CAH, they are often multifocal and bilateral, and are found at or around the mediastinum. All the lesions demonstrated strong enhancement on MR study. If testicular lesions are present in patients with CAH, US monitoring will be adequate for diagnostic evaluation.

Figure 1. T1 weighted MR image of a patient with CAH (top left); T2 weighted (top right); T1 weighted Gd enhancement image (bottom left); and subtraction image (bottom right).

References
5. Avila N.A et al., AJR, 172(4), 1002-6, 1999