ONE-STAGE PASSERINI-GLAZEL TOTAL REPAIR FOR PRIMARY GENITOPLASTY IN FEMALE CONGENITAL ADRENAL HYPERPLASIA: INTERMEDIATE OUTCOME IN 21 PATIENTS

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Introduction: Aims of primary-feminizing-genitoplasty (PFG) for congenital-adrenal-hyperplasia (CAH) in female are reduction of clitoris size maintaining vascularity and innervation (clitoroplasty), feminization of labioscrotal-folds (labioplasty), and division of uro-genital-sinus (UGS) obtaining normal placed urethra and vagina (vagino-urethroplasty). PFG could be performed very early in life as a complete repair in a single stage. Long-term follow-up of PFG for CAH in our Institutions (mainly performed following Hendren strategies) have shown need of secondary or redo-vaginoplasty in a significant number of patients. From 1999, we started using one-stage-Passerini-Glazel-total-repair in order to improve our cosmetic and functional results
Patients and Methods: One-stage-Passerini-Glazel-total-repair was our technique of choice. Magnitude of the procedure depends on degree of virilization and vaginal location. Lithotomy position and perineal approach were utilized in most of the patients. Knee-chest position and transanorectal approach and total UGS mobilization were reserved for the most difficult cases. Single or full-Passerini-Glazel-flaps were utilized in all the patients except four cases with absent or very short UGS. From 1999 to 2002, 28 patient affected by CAH underwent primary feminizing genitoplasty in our Institutions. Twenty-three patients were treated using simple-Passerini-Glazel-flap (UGS-mucosa only), 1 using full-Passerini-Glazel-flap (a tube made with UGS-mucosa and penile-skin), and 4 had only UGS distal cutback
Results: Intermediate outcome is available in 21 patients and was evaluated 1 year after the operation with voiding-cystourethrogram (VCUG) and EUA in 11 patients and with VCUG and outpatient-examination in 10. All 21 patients continue their regular follow-up at the Pediatric-Urology and Pediatric-Endocrinology Outpatient Clinics at our Institutions. In comparison with one stage or two stages surgical repairs previously in use in our Institutions, cosmetic and functional main improvements of Passerini-Glazel-technique are the short distance obtained between the clitoris and the urethral meatus and the well-separated openings of urethra and vagina (absence of residual UGS). Three patients showed a distal urethro-vaginal fistula. The mucosal bridge between urethral meatus and vaginal introitus, obtained with Passerini-Glazel flap, is usually flat and adequate, however, persistence of protruding mucosal bridge (reproducing an urethral/vaginal prolapse) was noticed in 4 patients
Conclusions: Female CAH is the most frequent form of ambiguous genitalia. One-stage Passerini-Glazel total repair (clitoroplasty, labioplasty, and vagino-urethroplasty) represents a great improvement in treatment of CAH. It allows one-stage and complete superb reconstruction of normal female genital anatomy. Passerini-Glazel UGS-mucosa-flap allows incomparable wide separation between urethral meatus and vaginal introitus. Passerini-Glazel procedure may achieve a significant decrease of number of vaginoplasty revisions at puberty
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