

46 XY Females with Male Karyotype: A Case Series in Malaysia

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ABSTRACT

Objectives: To compare the cases of phenotypic female patients that presented with male karyotype and the histopathological examination (HPE) results of their gonads. **Methods:** A retrospective review of five cases of 46 XY female patients that were managed by the Paediatric and Adolescent Gynaecology (PAG) unit, Department of Obstetrics and Gynaecology of UKM Medical Centre from 2009 to 2017, were conducted. **Results:** These patients, with female phenotypes, presented in adolescence or early adulthood with primary amenorrhoea with varying degrees of puberty. One was tall with breast development, another was very short with multiple co-morbidities, one had clitoromegaly and the other three had no secondary sexual characteristics. They were examined and had karyotyping and hormonal profiles done with ultrasound and MRI. Gonadectomies were performed once their 46 XY karyotype were confirmed. Histopathological examinations of their gonads were performed and the results varied from dysgenetic gonads to having testicular tissue to malignancy. All five had female gender identities and were given hormonal treatments and were counselled on fertility. One had a clitoroplasty performed. **Conclusion:** Female patients with 46 XY karyotypes require gonadectomies done at different timings depending on their diagnoses and gender identities. Risk of malignancy of their gonads need to be ascertained. Pubertal induction need to be managed. Issues such as fertility and gender identity need to be addressed. The managing team need to be sensitive in managing these rare cases as the diagnoses can be psychologically devastating.

KEY WORDS:

46 XY Female, Disorder of Sexual Development (DSD), Gonadal Dysgenesis, Swyer Syndrome, Androgen Insensitivity Syndrome, mixed gonadal dysgenesis, delayed puberty, paediatric and adolescent gynaecology

Ultrasonography and Clinical Outcomes following Surgical Anti-Incontinence Procedures (Monarc vs Miniarc): 5-year Review

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ABSTRACT

Objective: To evaluate the ultrasound morphology and its clinical outcome among women who had undergone MiniarcTM vs MonarcTM in the treatment of stress urinary incontinence (SUI), 5-year review. **Study Design:** This is a 5-year review study from a previous reported prospective study on 147 patients with USI and undergone either Miniarc or Monarc surgery. From March 2010 to December 2011, patients with clinically SUI and urodynamic stress incontinence (USI) were studied. Objective cure of SUI was defined as no urinary leakage on provocative filling cystometry and 1-hour pad test of <2 g. Subjective cure of SUI was the negative response to UDI-6. Introital ultrasound at 3-years explored the sling and bladder neck's position, mobility, sling tension, percentile of urethra where the sling was located and urethral kinking. **Results:** Postoperative data was available from 138 women. The ultrasound objective data for successful treatment post-operative follow-up was available from 123 women (51 Monarc, 72 Miniarc) at 3rd year, the rest failed to follow-up. A bladder perforation was diagnosed in the Monarc group and no exposure noted in both groups. At 3rd year, rest and during Valsalva, analogous distances of the bladder neck and sling as well as TU (distance between the mid position of the sling and center of urethral core) were similar for both procedures. In both groups, US (urethral shortest diameter) and UI (urethral longest diameter) were comparable at Valsalva that was significantly shorter and longer respectively compared to the values at rest. Both slings demonstrated caudally shift in the x-axis and y-axis. Miniarc position shifted more caudally on the x-axis (xt) at rest with concomitant more caudally shift on the y-axis on straining than Monarc. Percentage of urethral kinking remained similar. Failure rate increased with Miniarc but statistical analysis failed to detect any significant difference between the 2 groups with regards to the objective and subjective cure (Miniarc 87.8% and 82.9% vs Monarc 91.1% and 89.3%; p>0.05). **Conclusions:** In conclusion, Miniarc and Monarc maintained comparable subjective and objective clinical outcomes at 3-years minimum follow-up. Ultrasonography evaluation revealed more shift in the position of Miniarc caudally on x-axis at rest and y-axis during straining compared to Monarc. Majority of urethral impingement was still noted in the Miniarc group with concurrent higher MUCP (maximum urethral closure pressure). The differing shift may be related to sling anchoring mechanism.

KEY WORDS:

Anti-incontinence surgery, mid-urethral slings, outcomes, single incision sling, ultrasonography