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Diagnosis and management of complex reproductive outflow tract disorders in adolescents

Introduction: Outflow tract disorders in the PAG population are sufficiently rare for most general gynaecologists to have had experience and confidence in recognition and management of this highly complex cohort of young women, most of whom will need more than the anatomical correction, with considerable attention to their psychological wellbeing.

Methods: We retrospectively reviewed the medical records of patients with uterine, cervical and vaginal anomalies that presented to the PAG service from 2002 to 2017. The anomalies were classified according to the ESHRE/ESGE classification.

Results: A total of 102 cases of female congenital anomalies of the reproductive tract were identified from a cohort of 635 patients referred, of which 40 were obstructive anomalies. These conditions included patients with lower vaginal atresia managed by creation of surgical neovagina with pull through procedure (2), complete vaginal atresia with partial cervical atresia with the presence of a uterus (2) both requiring hysterectomy for life-threatening sepsis, triad of didelphys, obstructed hemi-vagina, and ipsilateral renal agenesis (17) requiring division of obstructed vaginal septum and in 3 of these cases hemi-hysterectomy for subsequent manifestation of unilateral cervical obstruction, unicornuate uteri with rudimentary functional cavity (2) requiring laparoscopic hemi-hysterectomy, unicornuate uteri with cervical and vaginal atresia requiring hysterectomy, and hypoplastic uteri with vaginal atresia (3) requiring hysterectomy. There were also transverse vaginal septa (6), imperforate hymen (6), bicornuate uterus with unicollis and transverse vaginal septum (1), and hypoplastic uterus with transverse vaginal septum (1) that were all managed with division of septa. Of the 102 patients, there were also 22 cases of mullerian tract agenesis (MRKH), in which there was an over 90% success rate from non-surgical neo-vagina creation, and 10 patients with bladder exstrophy or cloacal anomalies, of which three, had five pregnancies resulting in live late preterm babies. MRI is used to facilitate diagnosis in all cases of suspected congenital anomaly, and our extensive data on concordance and discordance with diagnosis demonstrates accuracy of MRI for diagnosis of uterine anomalies (94%), cervical anomalies (88%) and vaginal anomalies (78%), with an all structure concordance of 69%.

Conclusion: An understanding of these complex anomalies, which range from minor obstructions to major and life-threatening conditions, is required for appropriate recognition, assessment and management. Timely referral to specialist paediatric and adolescent gynaecologists for investigation, diagnosis, counselling, informed consent, and planning for surgery, would constitute optimal care for this complex cohort of young women.

Biography

Professor Rebecca Kimble is a Paediatric and Adolescent Gynaecologist (PAG) with 18 years' experience establishing and running the Queensland PAG service at the Quaternary Royal Brisbane and Women's and Lady Cilento Children's Hospitals. She has extensive experience with the management of complex congenital conditions of the female reproductive system, and over 25 years' experience with tertiary obstetrics in clinical and leadership positions. She is an Independent Health Advisor to the Australian Federal Government on Women's health. She has multiple publications in complex congenital conditions and is a member of the North American Society for Paediatric and Adolescent Gynaecology. She is medical lead Quality Improvement for the Queensland Government. She is founder and director of Queensland Clinical Guidelines, a globally utilized portal for evidence based Clinical Guidelines in Maternity and Neonatology. She chairs the Queensland Maternity and Neonatal Clinical Network and has achieved a reduction in unnecessary variation in clinical care.

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