# 46, XY Female—A Case Report

M K S Leow,* *MBBS, M Med (Int Med)*, K C Loh,** *FAMS, FACP, FRCPE*

## Abstract

***Introduction**: We examine a presumptive case of complete androgen insensitivity syndrome (CAIS) with certain unusual features. **Clinical Picture**: A woman with early onset osteoporosis gave a history of primary amenorrhoea and surgery for intraabdominal gonads. She subsequently defaulted follow-up and hormone replacement therapy. Endocrinological evaluation revealed hypergonadotrophic hypogonadism associated with a 46,XY karyotype. **Treatment**: Therapy included reinforcement of the female phenotype and oestrogen replacement. **Outcome**: There was gradual development of her secondary sexual characteristics and improvement in her bone mineral density. **Conclusion**: Patients with CAIS need proper counselling and education according to their psychosexual make-up and sociocultural factors. The importance of long-term oestrogen replacement in a young subject post-gonadectomy cannot be overemphasised as illustrated in our case.*

***Ann Acad Med Singapore* 2001; 30:71-5**

***Key words: Androgen insensitivity syndrome, Atypical presentation, Male pseudohermaphroditism, Management, Testicular feminisation syndrome***

* *Registrar in Endocrinology*
** *Consultant Physician and Endocrinologist*
*Endocrine Unit*
*Department of General Medicine*
*Tan Tock Seng Hospital*
*Address for Reprints: Dr Melvin Leow Khee Shing, Department of Medicine, Tan Tock Seng Hospital, 11 Jalan Tan Tock Seng, Singapore 308433.*