GnRH-analogue treatment in children with congenital adrenal hyperplasia (CAH): data from a multicenter CAH registry

F. Schreiner[1], J. Rohayem[2], S. Fricke-Otto[3], S. Golembowski[4], N. Jorch[5], K.O. Schwab[6], K. Warncke[7], U. Zanier[8], on behalf of the German CAH registry (DGKED-QS /AGS)

[1] Pediatric Endocrinology, Children's Hospital, University of Bonn, Germany
[2] Center of Reproductive Medicine and Andrology, Department of Clinical Andrology, University of Münster, Germany
[3] Pediatric Endocrinology, HELIOS Klinikum Krefeld, Germany
[4] Pediatric Endocrinology, Sana Klinikum Lichtenberg, Berlin, Germany
[5] Pediatric Endocrinology, Evangelisches Klinikum Bethel, Bielefeld, Germany
[6] Pediatric Endocrinology, Children's Hospital, University of Freiburg, Germany
[7] Pediatric Endocrinology, Children's Hospital, Klinikum Schwabing, TU Munich, Germany
[8] Pediatric Endocrinology, Krankenhaus Dornbirn, Austria

INTRODUCTION

Final height in CAH patients is generally assumed to be lower than the population norm. Besides CAH subtype and age at diagnosis, timing of pubertal development is considered to have a significant impact on final height. In most CAH patients, puberty starts within normal ranges, although at a somewhat earlier mean age compared to reference populations. CAH-complicating gonadotropin-dependent precocious puberty has been reported in few cases, especially in conditions of late CAH-diagnosis and treatment initiation, and often requires additional treatment with GnRH-analogues.

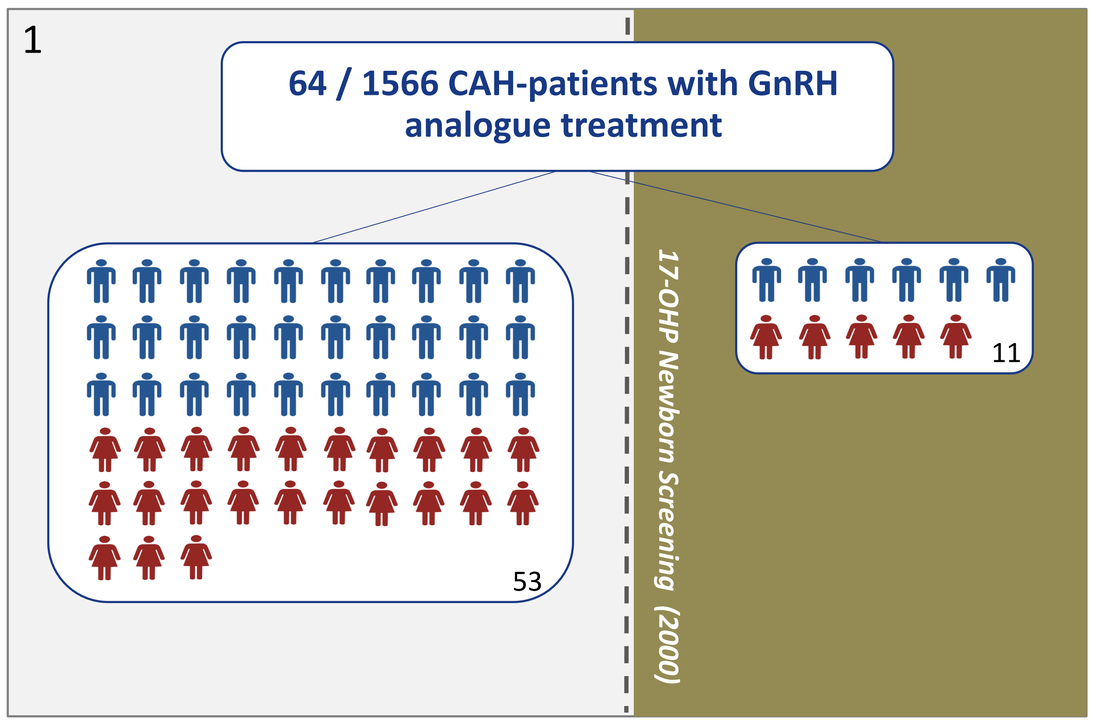
PATIENTS & METHODS

We retrospectively assessed frequency, clinical parameters and height outcome of GnRH-analogue treated CAH-patients from the German CAH registry (DGKED-QS/AGS), comprising longitudinal data of a total of >1500 CAH-patients.

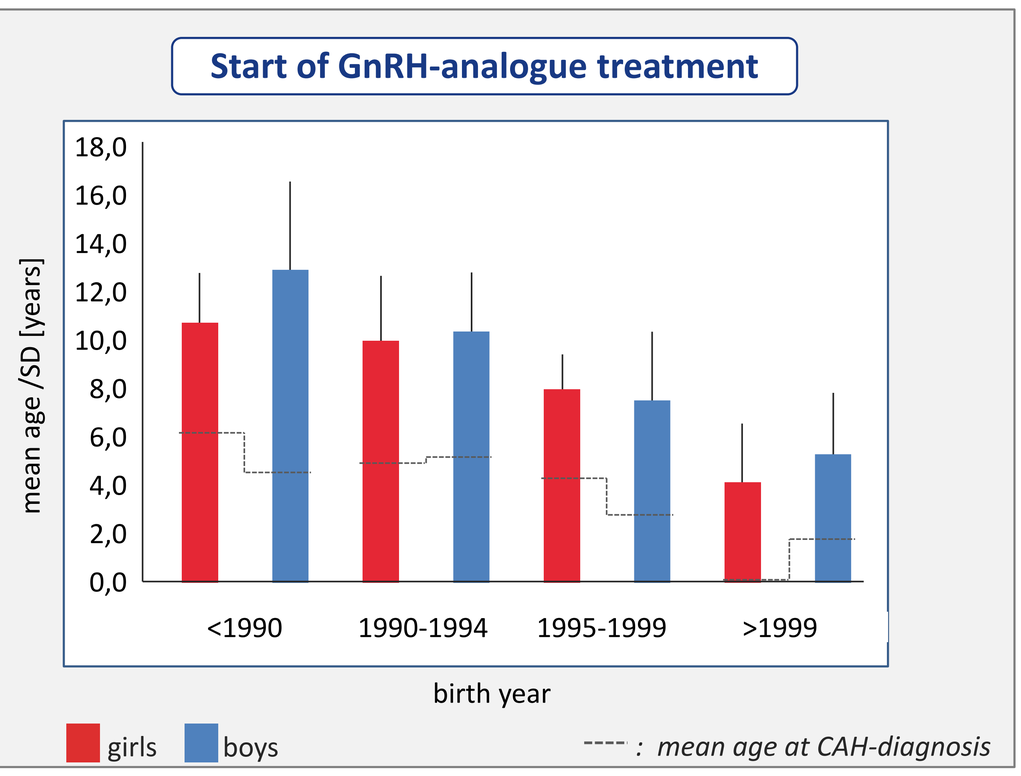
RESULTS

- The majority of 64 GnRH-analogue-treated patients were born in the pre-newbornscreening era (***Fig. 1***).
- Mean age of CAH-diagnosis decreased over time, even within the pre-newbornscreening era (***Fig. 2***, *dotted lines*).
- Only one third (37,5 %) of them suffered from salt-wasting CAH (***Fig. 3***).
- Mean age of first database entry of GnRH-analogue treatment was 8.81 /9.24 yrs (girls /boys), with a significant decline from the 1980s to the 2000s (***Fig. 2***).
- Some children without clinical signs of gonadarche received GnRH-analogue treatment, (presumably) solely initiated for auxological reasons (***Fig. 4***).
- Mean height-SDS at GnRH-analogue treatment start was +1.18 SDS (+1.74 SDS TH-corr.) in girls /+ 0.62 (+1.13) SDS in boys, with accelerated bone age (+3.29 /+3.35 yrs) in both genders. At transition, girls reached a normal final height (-0.15 SDS, TH-corr.), while most boys remained subnormal (***Fig. 5***).

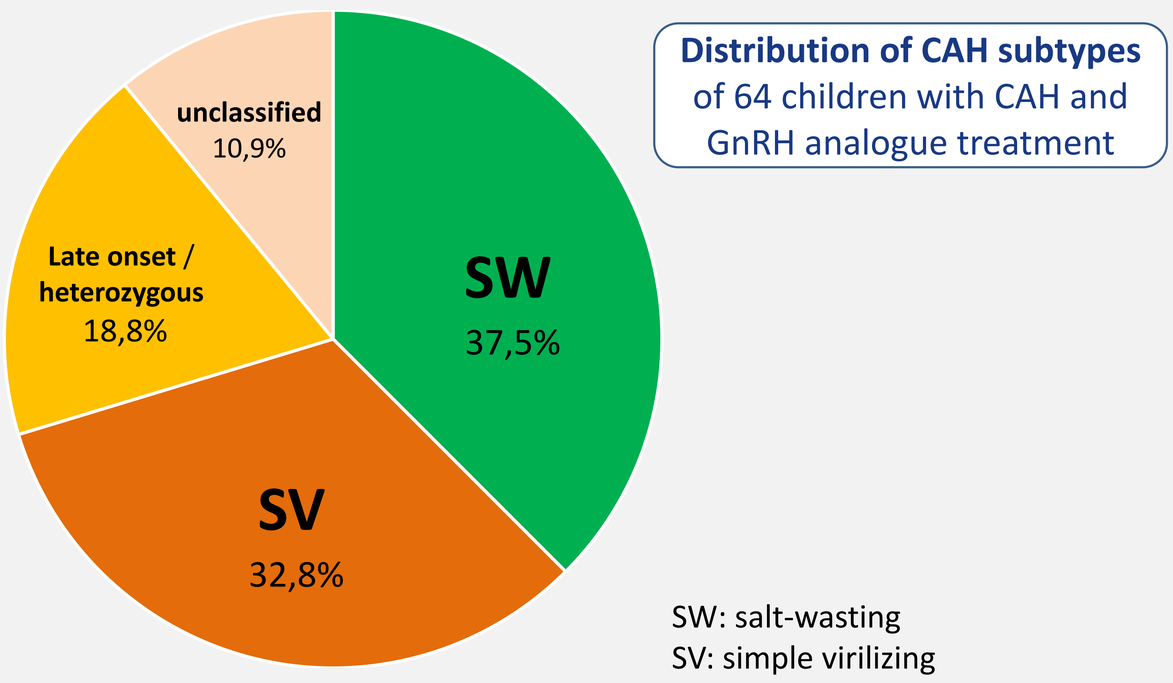
1



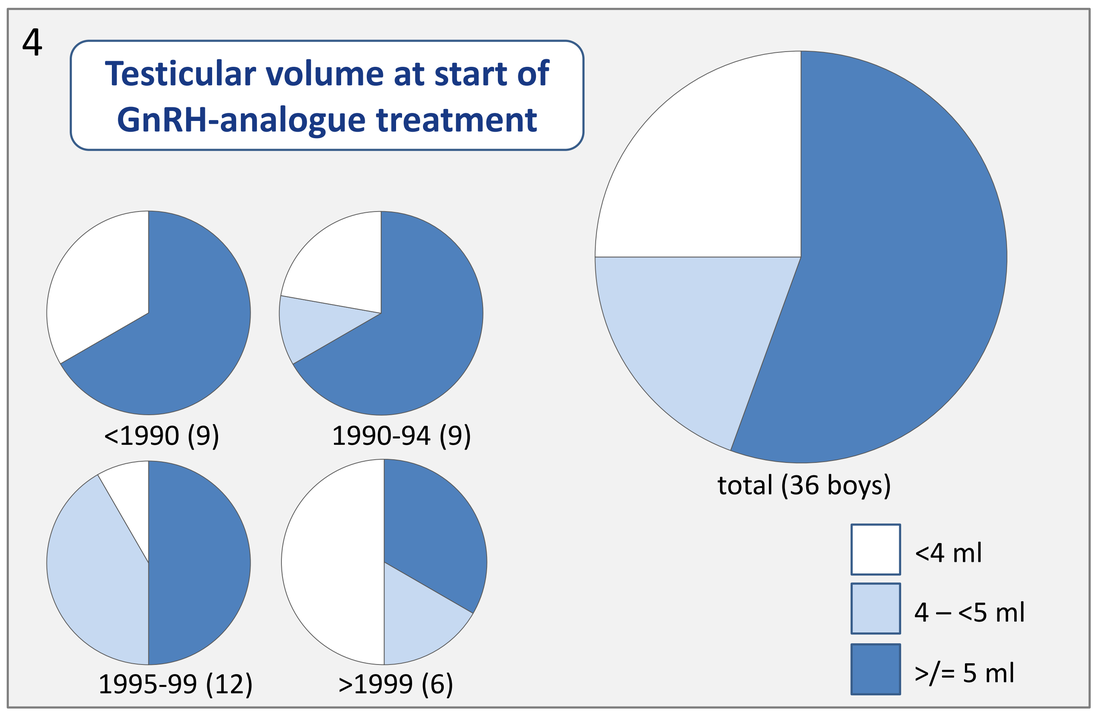
2



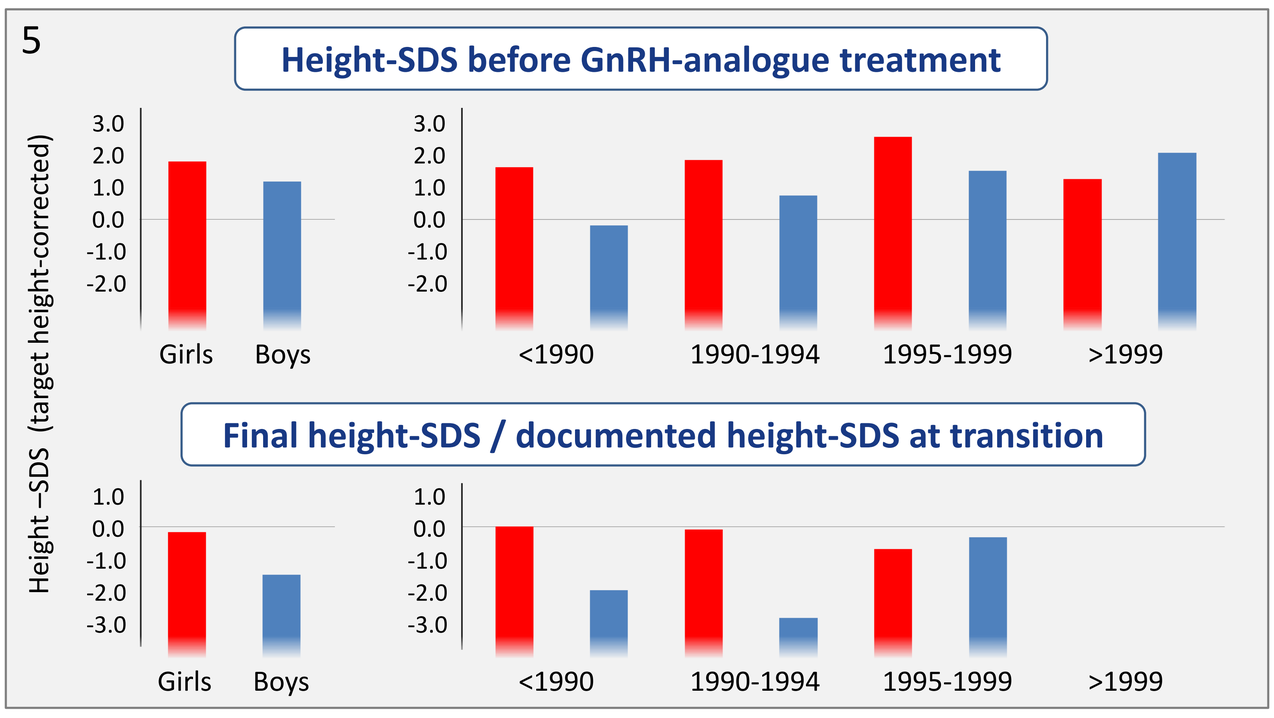
3



4



5



CONCLUSIONS

- GnRH-analogue treatment in children with CAH is rare, especially in the 17-OHP newborn screening era.
- A decline of mean age at CAH-diagnosis paralleld by decreasing use of GnRH-analogue treatment in recent decades supports the concept that chronic hyperandrogenemia (or its cessation) can trigger central precocious puberty.
- The auxological data of this cohort analysis seems to indicate that GnRH-analogue treatment may have a beneficial effect on final height, especially in girls.