Determining Prevalence of Renal Disease in an Australian Sickle Cell Disease Cohort

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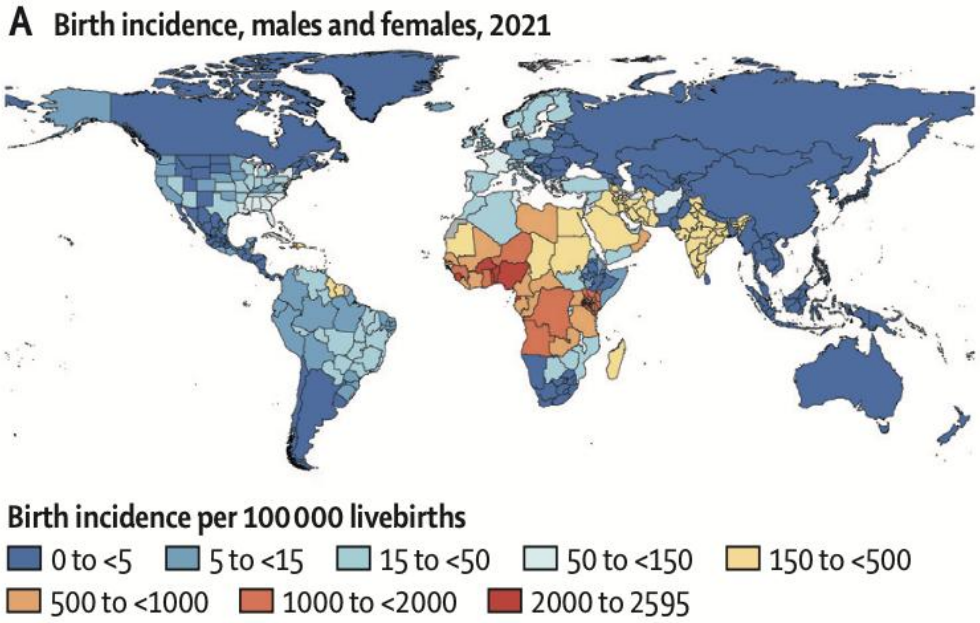
Introduction

- Autosomal recessive disorder
- Hb S
- Vaso-occlusive disease and haemolytic anaemia
  - Sickle cell nephropathy
    - CKD and ESKD
- Geographic variation



Progression to CKD

- The renal complications of SCD include
  - Chronic glomerular damage
  - Vascular damage
  - Chronic tubular damage
  - Renal medullary carcinoma
  - Recurrent AKIs due to recurrent sepsis related to functional asplenism

- Due to these renal complications, CKD occurs at a much earlier age than the general population

- The presence of sickle cell nephropathy is associated with increased mortality

- Patients that progress to ESKD have a 3x higher mortality rate compared to the general dialysis population

Prevention of sickle cell nephropathy

- Key preventative strategies
  - Recognition of hyperfiltration and subsequent over-estimation of eGFR
  - Annual screening for albuminuria from age 10
  - Annual screening for haematuria – renal medullary carcinoma
  - Imaging renal tract
  - Early referral to nephrologist

  - Disease modifying therapies to treat underlying SCD
    - Hydroxyurea – prevents vaso-occlusive disease and has been associated with lower rates of albuminuria[1]
    - Red cell exchange
    - Allogenic stem cell transplant

1. Laurin LP, Nachman PH, Desai PC, Ataga KI, Derebail VK. Hydroxyurea is associated with lower prevalence of albuminuria in adults with sickle cell disease. Nephrology Dialysis Transplantation. 2014 Jun 1;29(6):1211-8.

Methods

- Single-centre, prospective cohort study in which demographic, clinical, biochemical and quality of life data were collected

- 20 patients consented

Results

Demographic and clinical characteristics of patients with sickle cell disease	
Characteristic	Cohort, *n* (%)
Total	20
Sex	
Female	8 (40)
Male	12 (60)
Median age	39.5 (IQR 29 - 47.25)
Blood pressure	
Systolic blood pressure (mmHg), mean	$_{1}$22mmHg (SD 116-127)
Diastolic blood pressure (mmHg), mean	$_{7}$1mmHg (SD 67-75)
Prior sickle cell crisis documented	17 (85)
Management	
Hydroxyurea	12 (60)
Red cell exchange	19 (95)

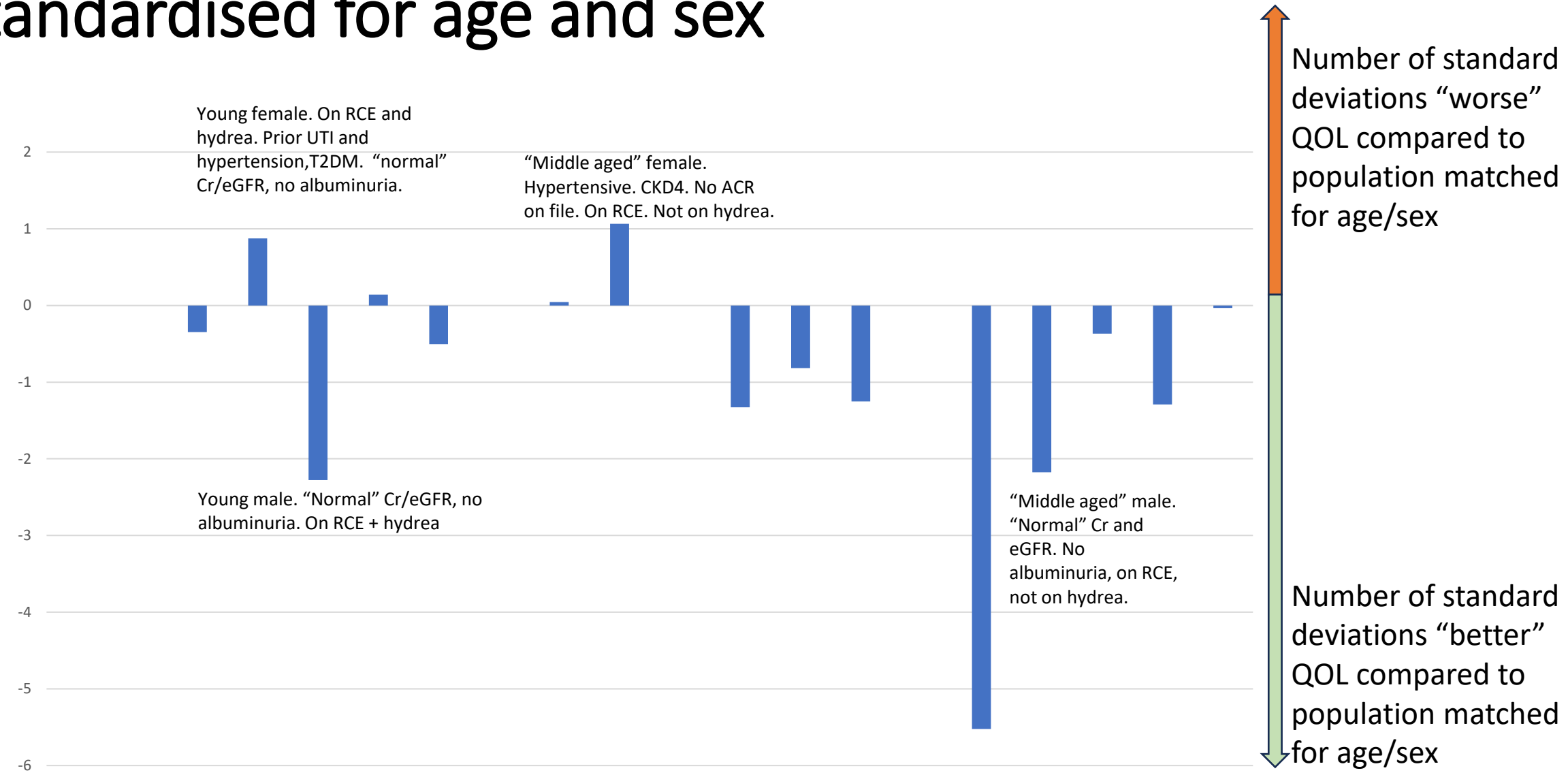
Results

Biochemical and imaging findings of patients with sickle cell disease	
Outcome	Cohort, *n* (%)
Total cohort	20
Blood results available	19 (95)
Serum creatinine (mmol/L), mean	71 (95% CI 49-92)
eGFR > 90	16 (84.2)
Urine results available	16 (80)
Red cell count > 10	0
White cell count > 10	0
Positive culture	1 (6.3)
Urine ACR abnormal	6 (37.5)
Range of abnormal results (mg/mmol)	5.2 - 135
Urine PCR abnormal	4 (25)
Range of abnormal results (mg/mmol)	6-192
Renal imaging performed last 5 years	14 (70)
USS	8 (57.1)
CT	6 (42.9)
Abnormality detected	4 (28.6)
Completed all screening within last 12 months	11 (55)

Results

Renal complications of sickle cell disease	
Outcome	Cohort, *n* (%)
Total cohort	20
History of renal event	5 (25)
AKI	4 (20)
Haematuria	5 (25)
Urine infection	2 (10)
Seen in renal clinic	4 (20)

Quality of life data – AQoL-8D Z scores (n=15) standardised for age and sex



Conclusion

- This SCD cohort had elevated rates of renal events and albuminuria despite their relatively young age (mean age 39.5)

- Routine screening was frequently incomplete, and 40% were not receiving hydroxyurea.

- There were inconsistent QoL data between surveys, however there was a correlation between renal disease and poorer QoL indices.

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