ARM in ARM?

Birmingham Women's and Children's NHS Foundation Trust

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Investigating the co-occurrence of anorectal malformations and labioscrotal anomalies

Purpose

Anorectal malformations (ARMs) are often associated with other congenital defects:
⟶ No evidence addressing the co-occurrence of ARMs with **labioscrotal anomalies (LSAs)**;
⟶ A **systematic review** of the literature was performed to analyse the co-occurrence of ARMs and LSAs for patterns of association.
Further aim: bring clarity to the complicated matter of **nomenclature** relating to ARMs and LSAs.

Methods

'Low' ARM
- Perineal (cutaneous) fistula
- Vestibular fistula
- Rectovaginal fistula
- Anal stenosis

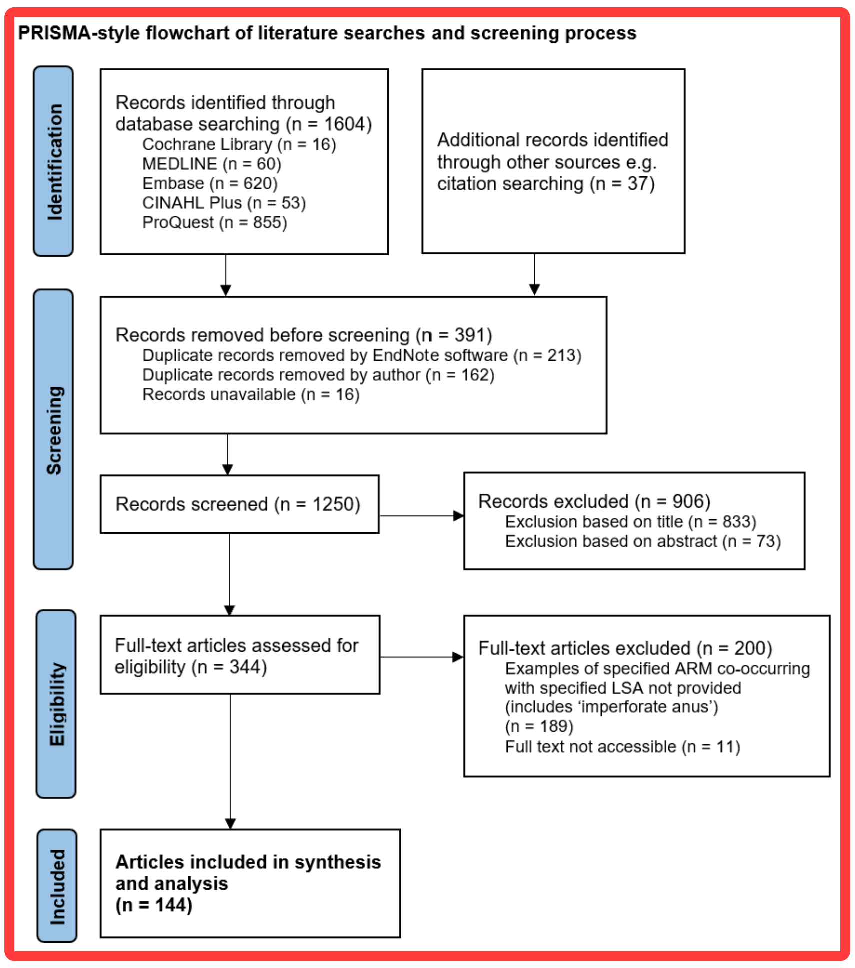
'High' ARM
- Rectourethral prostatic fistula
- Rectourethral bulbar fistula
- Rectobladder neck fistula
- Rectovesical fistula
- Rectal atresia

Complex ARM
- Cloaca
- No fistula
- Pouch colon
- H-type fistula

Labioscrotal Anomalies
- Bifid scrotum
- Ectopic scrotum
- Accessory scrotum
- Hemiscrotum
- Penoscrotal fusion
- (Peno)scrotal transposition
- Scrotal raphe anomaly
- Scrotoschisis
- Scrotal agenesis
- Hemiscrotal agenesis
- Proximal hypospadias
- Labial hypoplasia
- Labial hypertrophy
- Labial fusion
- Accessory labial fold
- Other

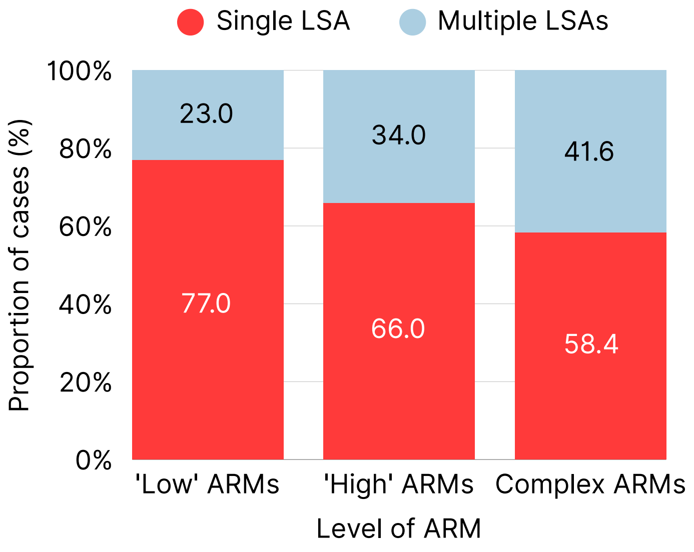
● Classified as "Major clinical groups" in the Krickenbeck classification
● Classified as "Rare/regional variants" in the Krickenbeck classification

Both the number of individuals with at least one ARM-LSA co-occurrence and the total number of LSAs associated with each type of ARM were recorded.



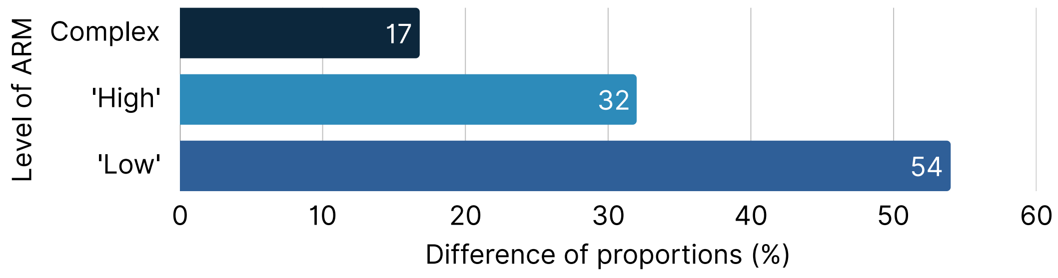
Results

Proportion of ARM cases associated with single vs multiple LSAs ('low', 'high', complex ARMs)



- **144 papers** met inclusion criteria, reporting co-occurrence of ARMs with LSAs (**319 patients**).
- Patients with complex ARMs had proportionally the most LSAs associated with their ARM.
- The absence of data reporting prevalence of each type of LSA in isolation (i.e., without an ARM) limits the potential to associate specific LSAs with specific ARMs.

Difference between proportions of ARM cases with single vs multiple LSAs



Conclusion

This systematic review indicates that **more severe ARMs are associated with a greater number of LSAs.**
A **significant proportion of 'low' (i.e., less severe) ARMs may co-occur alongside at least one LSA.**
This study emphasises the need to **establish a close collaboration between paediatric surgeon and urologist**, ideally in a joint clinic setting, in the management of patients with any type of ARM, not only for renal but also likely LSA related comorbidities.

SCAN for

- Abstract
- Bibliography
- List of included studies

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