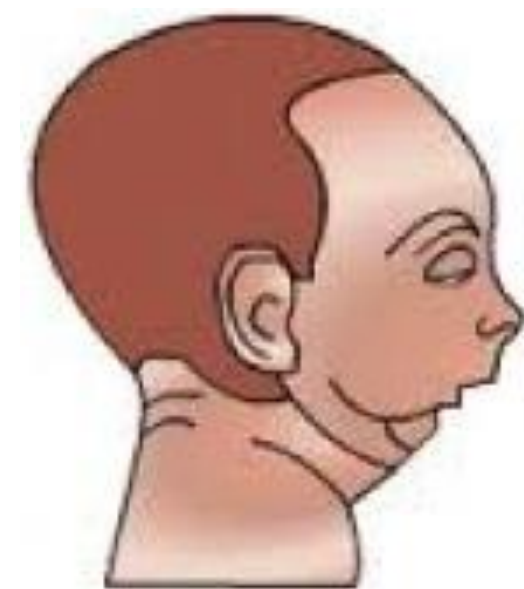


1. Introduction

Robin sequence (RS) is a congenital condition which occurs in 1 in 24,500 babies per year on average (Paes et al. 2015). The condition has three main features:



- Micrognathia**
 - A noticeably smaller jaw
- Glossoptosis**
 - A tongue further back in the mouth (due to micrognathia)
- Upper airway obstruction**
 - Airway obstruction and breathing difficulties (due to glossoptosis)

Figure 1: An illustration of a child with RS.

The features of RS can cause feeding and breathing difficulties from birth. Additionally, children with RS are often born with cleft palate, resulting in speech difficulties characterised by velopharyngeal insufficiency and/or cleft speech characteristics.

- ❑ **Velopharyngeal insufficiency (VPI)** - Air entering and leaking through the nose due to structural malformation resulting in hypernasal speech and nasal airflow errors.
- ❑ **Cleft speech characteristics (CSCs)** - Types of articulation errors commonly heard in children with cleft palate. CSCs are labelled as anterior, posterior or non-oral and increase in terms of severity. CSC types can co-occur.

Existing Research

Children with **RS and cleft palate (CPRS)** are thought to have increased speech and language difficulties compared to children with **isolated cleft palate (ICP)**. However, current research investigating speech and language outcomes for this population is limited:

- ❑ CSCs are more frequent and severe in children with CPRS than in children with ICP at age 5 years (Hardwicke et al, 2016).
- ❑ Mixed results are reported for VPI when comparing children with CPRS to those with ICP (Hardwicke et al., 2016 and Stransky et al., 2012).
- ❑ Language is under researched. Children with RS are thought to have language delay (Thouvenin et al., 2013; Williams et al., 1981).

Children with CPRS are currently on the same treatment pathway as children with ICP in the UK. **Further research is necessary** to better understand the treatment and management that children with CPRS require for speech and language development.

2. Aims

Aim: To investigate whether children with cleft palate and Robin Sequence (CPRS) have different speech and language outcomes from children with isolated cleft palate (ICP).

- ❑ At 5 years, do children with CPRS have increased VPI?
- ❑ At 5 years, do children with CPRS have increased levels of CSCs?
- ❑ Do children with CPRS have poorer expressive and receptive language outcomes than children with ICP at 3 years?

3. Method

- ❑ **Design:** Retrospective data collection within the Northern Regional Cleft Lip and Palate Service in Newcastle Upon Tyne. Full Caldicott approval granted.
- ❑ **Participants:** Children born between 01/01/2004 to 31/12/2013 with ICP or CPRS. A total of 74 children, 37 with CPRS and 37 with ICP, were matched by socio-economic status, gender, date of birth and age at 5-year speech audit.
- ❑ **Process:** Information was gathered from case files on early feeding, early airway management, cleft palate type, secondary surgery and presence of fistula after primary surgery. Speech and language assessment results at age 3 years and 5 years provided data for rates of velopharyngeal insufficiency and cleft speech characteristics.

4. Results and Discussion

Speech outcomes

Cleft speech characteristics (CSCs)

Children with CPRS were found to have significantly more frequent and more severe CSCs than children with ICP at 5 years ($p=0.002$). This is consistent with current literature (Hardwicke et al, 2016).

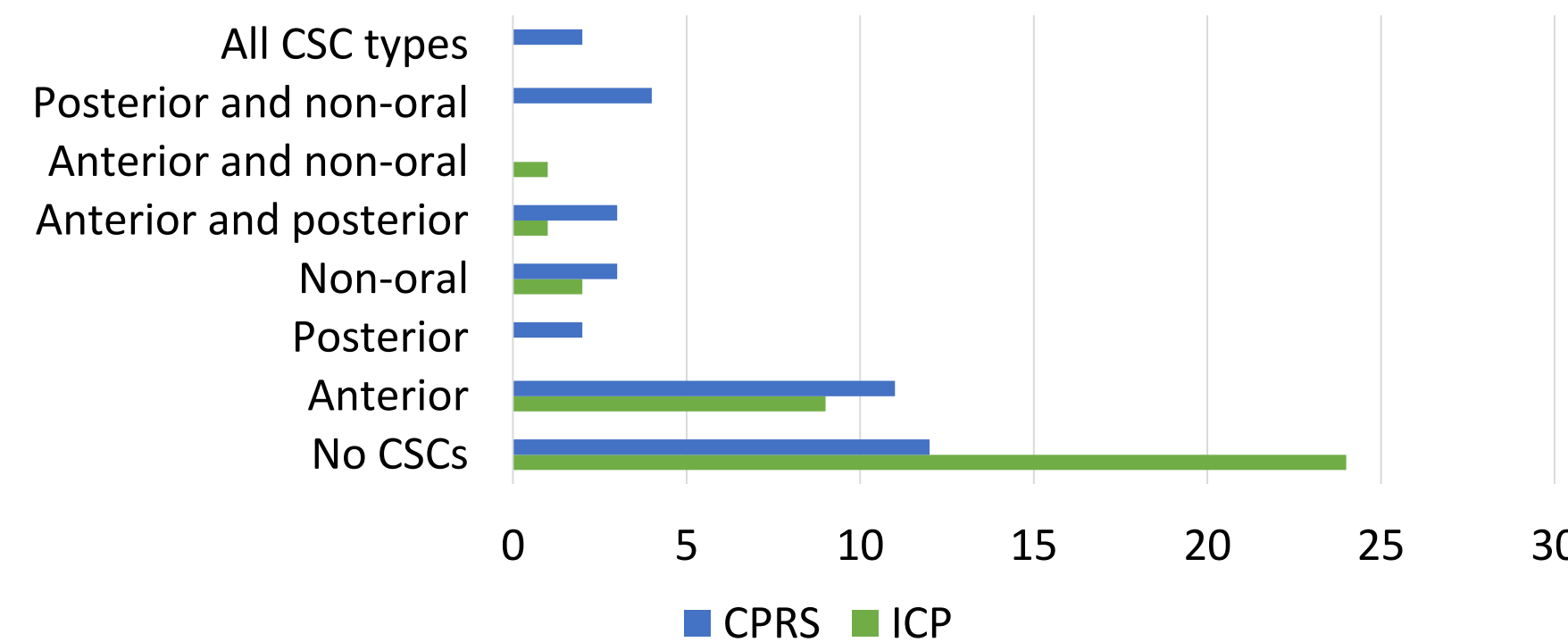


Figure 2: A bar chart showing the CSC types found across participants in each group.

Velopharyngeal insufficiency (VPI)

More children in the CPRS group had VPI at 5 years but this was not significant ($p=0.055$).

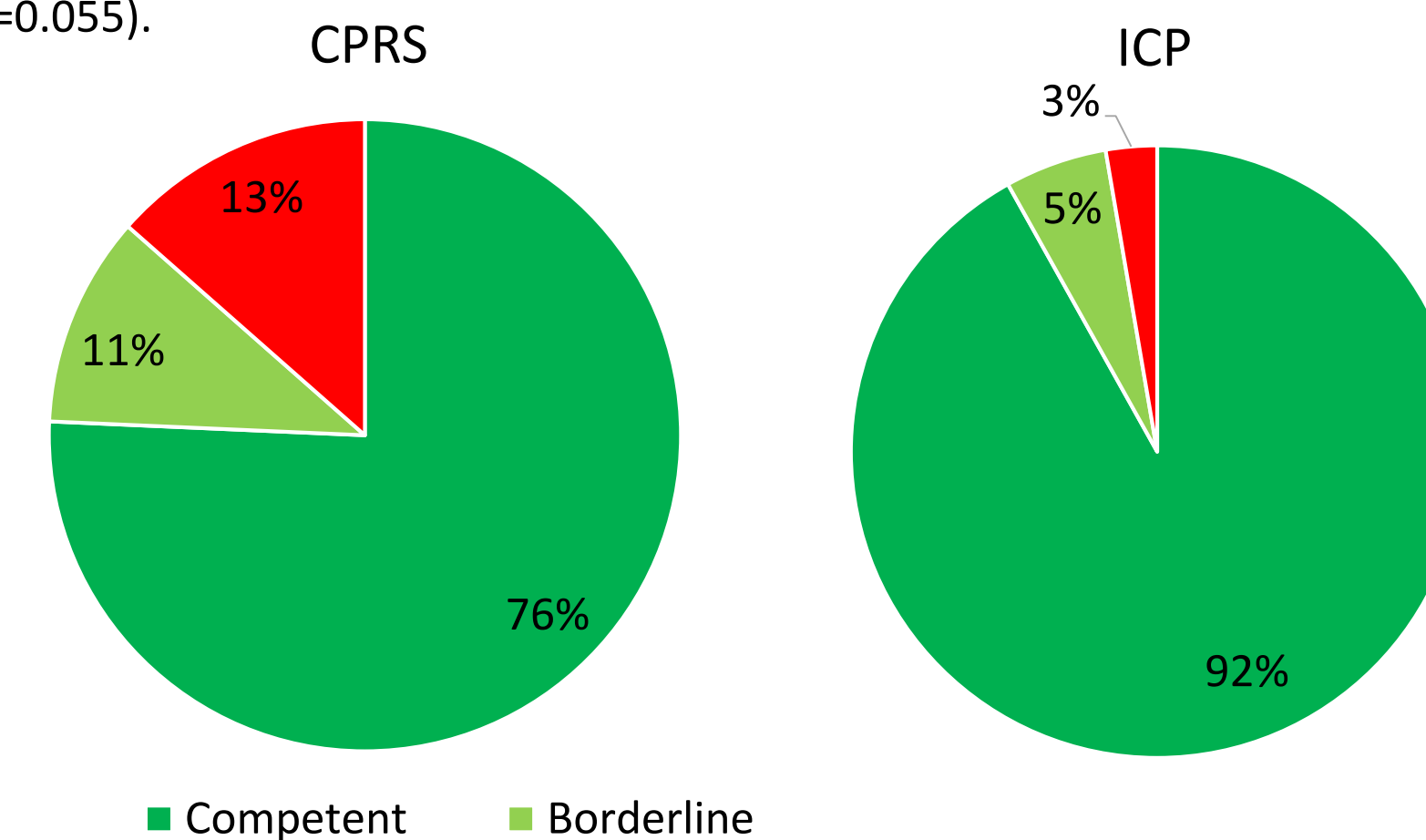


Figure 3: The percentage of participants with VPI graded as competent (no VPI), borderline (mild VPI) and incompetent (severe VPI).

Language outcomes

- ❑ No significant difference was found between the CPRS and ICP groups for receptive language ($p=0.326$) and expressive language ($p=0.496$) at 3 years.
- ❑ Outcomes contrast with previous research (Williams et al, 1981; Thouvenin et al, 2013). This could be due to the brief nature of the language screen used within the service.

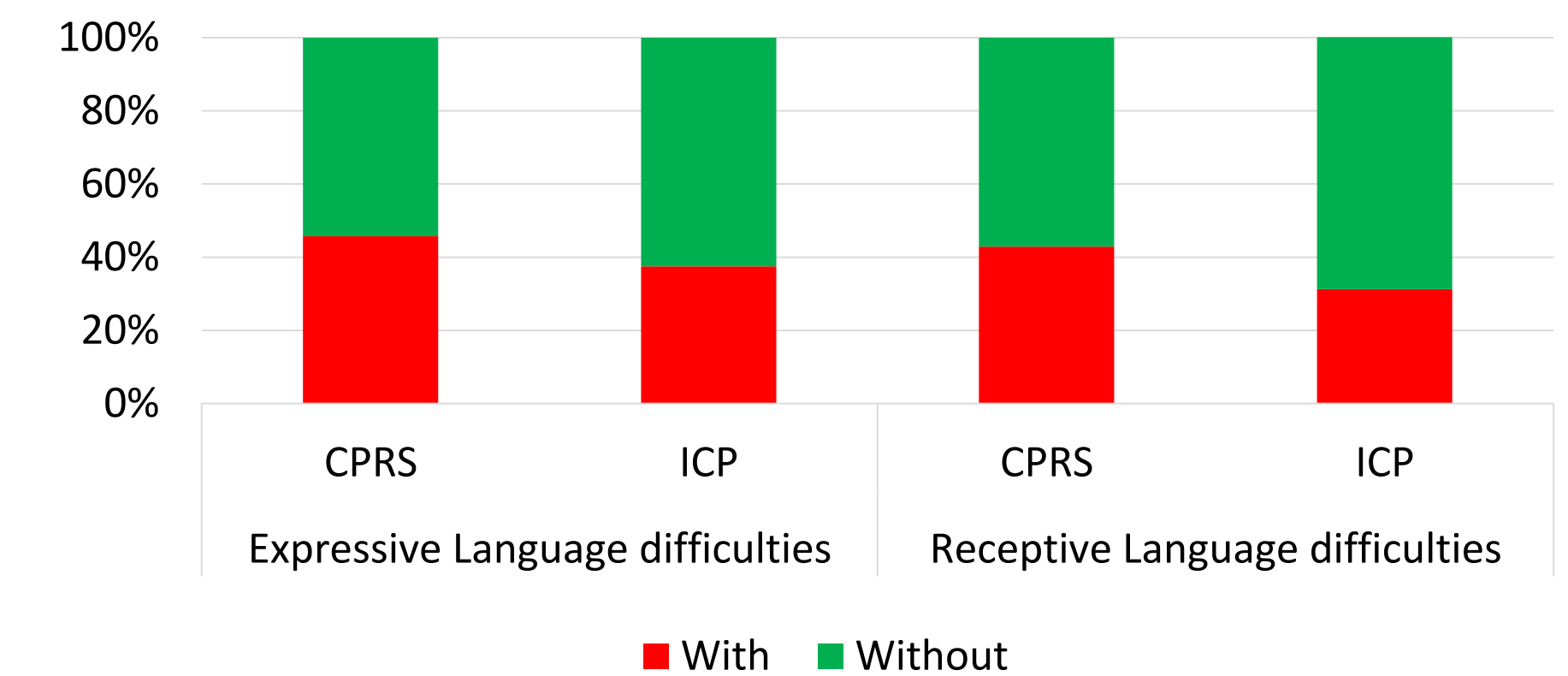


Figure 4: The percentage of participants in each group with and without expressive and receptive language difficulties.

Study limitations

- ❑ The retrospective nature of the study meant patients were excluded because their 5-year speech audit was missing or incomplete.
- ❑ Data was often missing due to restricted speech and language outputs during speech assessments, suggesting these children may have experienced speech and/or language delay which could not be accounted for in the results.

5. Conclusions

Increased CSCs in the CPRS group

- Children with CPRS experience **significantly more frequent and severe cleft speech characteristics** than children with ICP. Almost twice as many children in the ICP group presented with age appropriate articulation.

No significant difference in language

- Language outcomes were not significantly different. However, **further investigation is warranted** using more sensitive language assessments and with a larger cohort.

CPRS requires increased support

- This research, along with other studies, highlights the severity of cleft speech characteristics in children with Robin Sequence, suggesting children with CPRS may **require more support** than children with ICP.

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