

The benefits of universal newborn
hearing screening for permanent
childhood hearing impairment at teen
age

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Bilateral Permanent Childhood Hearing Impairment - PCHI



Hearing loss that is:

- Bilateral
- Permanent
- Congenital
- Moderate-profound
i.e. >40 dB HL

1.12 per 1000 live births (Davis et al., 1997)

Over 8,000 cases of PCHI identified in the UK
by newborn screening in 5 yrs 2008-13

$>43\%$ of all cases per yr of all 29 conditions in the USA
for which newborn screening is in place

- Sensitive periods for language development
- Unidentified PCHI places a child at significant risk of speech and language disorder and delay.



- 90% born to hearing parents
- Over 50% have no known risk factors

Need to identify these babies so that early intervention can be provided, e.g. sign language, hearing aids, CIs



Two stage Universal Newborn Hearing Screening (UNHS)

1. Oto-acoustic emissions



2. Automated ABR



SCIENCEPHOTOLIBRARY

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HOT Project
Hearing Outcomes in Teenagers

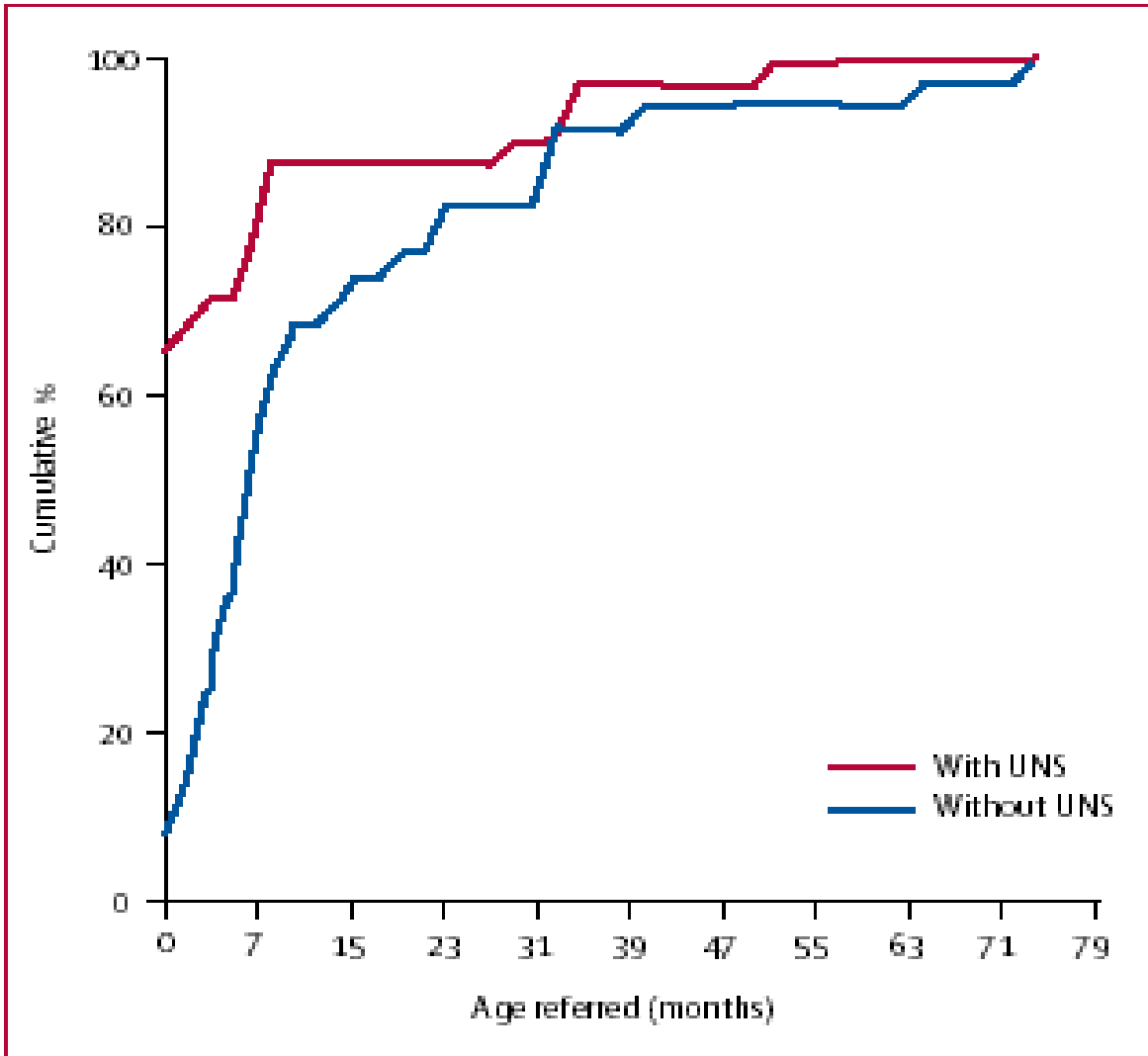
Phase 1: The Wessex Trial of UNHS

(October 1993 - October 1996)

| Testing Period | | Team 1 Hospital | | Team 2 Hospital | |
|----------------|-------------------|--------------------|------|--------------------|------|
| designation | duration (months) | A | C | B | D |
| 1 | 4 | 1586 | 1819 | 1071 | 1285 |
| 1 | 4 | 1664 | | 1150 | |
| 2 | 4 | 1547 | 1745 | 1149 | 1022 |
| 2 | 4 | 1547 | | 1149 | |
| 3 | 4 | 2264 | 1963 | 1591 | 1222 |
| 3 | 4 | 2264 | | 1591 | |
| 4 | 6 | | 2873 | | 1658 |
| 4 | 6 | | | | |

Numbers shown are total in-patient live births during periods **with** newborn screening (n=25,609).
 Total numbers of in-patient live births during periods **without** newborn screening (n=28,172) are not shown

cumulative percentage of all known cases of bilateral PCHI ≥ 40 dB in the population at age 7–9 years by birth in periods with and without universal newborn screening



Kennedy C, McCann D, Campbell J,
Kimm L, Thornton R.
The Lancet 2005, 366: 660-62

Phase 1: The Wessex Trial



- Controlled Trial of Universal Newborn Hearing Screening in 1993-96 birth cohort of 53,000
- Demonstrated that UNHS leads to earlier referral of PCHI

Kennedy et al *The Lancet* 2005, 366: 660-662

Next Question, addressed in Phases 2 & 3:

Is UNHS and early identification of PCHI associated with improved language outcomes later in life?

review of case for introduction of UNS

- UK government announces on 22nd June 2000, following national screening committee recommendation, that UNS will be introduced in 20 site pilot and rolled out to all UK districts over 3 years.

but, in North America....

- ‘better evidence about the effectiveness of UNS is needed and could be obtained via ...inception cohorts’
Thompson et al, *JAMA* 2001
- ‘the U.S. Preventive Services Task Force concludes the evidence is insufficient to recommend for or against routine screening of newborns for hearing loss during the postpartum hospitalization’

Nine years later

Phase 2: The Hearing Outcomes Project



- 2002-2004; children now 6-10 years old
- Wessex Trial sample combined with Greater London sample, both population-based

- 120 children with PCHI
 - 61 UNHS + 59 no UNHS
 - 57 confirmed \leq 9 months + 63 confirmed $>$ 9 months
- 63 children with normal hearing

Phase 2: The Hearing Outcomes Project

| | Born in periods with UNHS | Confirmed at ≤ 9 months |
|-------------------------|---------------------------|------------------------------|
| • Reading comprehension | superior | superior |
| • Expressive language | superior | superior |
| • Receptive language | superior | superior |

Kennedy et al. *New England Journal of Medicine* 2006, 354: 2131

McCann et al. *Archives of Disease in Childhood* 2009, 94: 293

Phase 2: the Hearing Outcomes Project

United States Preventative Services Task Force

2001: UNHS 'Insufficient Evidence'

2008: UNHS 'Recommend'



Another 9 years later.....Phase 3: The Hearing Outcomes in Teenagers HOT Project

Have the previously observed benefits to reading and language outcomes associated with early confirmation of hearing impairment continued into the teenage years?



| | Eligible phase 2 sample | Recruited phase 3 sample |
|-------------------------------|----------------------------|-----------------------------|
| Teenagers with PCHI | 120 | 76 (63%) |
| Teenagers with normal hearing | 63 | 38 (59%) |

Phase 3 outcome measures

**Pre-specified primary outcome measure =
York Assessment of Reading Comprehension (YARC)**

Other principal outcome was Receptive Language

- British Picture Vocabulary Scale (BPVS)
- Test for Reception of Grammar (TROG)

Non-verbal ability

- Ravens progressive matrices

All assessments undertaken in participants' homes

Hearing Outcomes in Teenagers Project

Other outcomes...

Theory of Mind

Behaviour

Quality of life

Health

Educational performance

Social functioning

Economic costs

Participants in phases 2 and 3

| | Phase 2 sample (n=120) | Phase 3 sample (n=76) |
|-------------------------------------|---------------------------|--------------------------|
| Gender (% male) | 56 | 51 |
| Mean(SD) age at phase 2 assessment | 7.9 (1.3) | 7.9 (1.1) |
| Mean(SD) non-verbal ability z-score | -0.71 (1.2) | -0.62 (1.2) |
| Severity of Hearing Loss | | |
| % Moderate | 52 | 50 |
| % Severe/Profound | 48 | 50 |
| Maternal Education Level | | |
| % No qualifications/ < 5 O Levels | 36 | 32 |
| % > 5 O Levels/ Some A Levels | 52 | 53 |
| % University degree or higher | 12 | 16 |

Phase 3 participants

| | Confirmation \leq 9 months (n = 35) | Confirmation $>$ 9 months (n = 41) |
|------------------------------------|---------------------------------------|------------------------------------|
| Gender (% male) | 54 | 49 |
| Mean (SD) age at HOT assessment | 16.8 (1.5) | 17.3 (1.3) |
| Mean (SD) NV ability z score | -0.26 (0.9) | -0.28 (0.8) |
| Severity of Hearing Loss | | |
| % Moderate | 45 | 42 |
| % Severe/Profound | 56 | 58 |
| English First Language at Home (%) | 97 | 88 |
| Cochlear Implants (% implanted) | 20 | 19 |
| Maternal Education Level | | |
| % No qualifications/ < 5 O Levels | 26 | 24 |
| % > 5 O Levels/ Some A Levels | 48 | 52 |
| % University degree or higher | 26 | 24 |

Birth in UNHS and early confirmation in phase 3 participants (n = 76)

| | UNHS (n = 37) | No UNHS (n = 39) |
|---|------------------|---------------------|
| Confirmation \leq 9 months (n = 35) | 24 | 11 |
| Confirmation $>$ 9 months (n = 41) | 13 | 28 |

YARC

see

look

play

journey

caught

tongue

haemorrhage

endogenous

paediatrician

Reading accuracy score = Total number of words
read correctly

YARC

It was the first day of Ryan's family holiday. They were staying in a cottage which overlooked the harbour in Peele Bay. It was a glorious sunny day, so the family had wandered down to the beach. Dad volunteered to look after their bags. Mum explored the beach, then joined Ryan and his sister in the foaming waves. Dad relaxed and read his magazine. When Mum had had enough of the water, she returned to sit with Dad. He had fallen asleep and was scarlet. She glanced around and realised her handbag was missing. It must have been stolen. Mum was furious with dad. Everyone hurried to the police station. Much to their surprise it had already been handed in and nothing was missing. The policeman said an old lady had found it in the beach toilets. Then mum remembered; she had left it there. Mum apologised to Dad and bought him a huge ice-cream.

- Where did Ryan and his family go to on holiday?
- How do you know that the cottage was close to the beach?
- Why did the family decide to go to the beach?
- In the first paragraph what does 'volunteered' mean?
- Why did Mum blame Dad?

Can you provide a short summary of the passage, making clear what the main events are?

YARC

It was the first day of Ryan's family holiday. They were staying in a cottage which overlooked the harbour in Peele Bay. It was a glorious sunny day, so the family had wandered down to the beach. Dad volunteered to look after their bags. Mum explored the beach, then joined Ryan and his sister in the foaming waves. Dad relaxed and read his magazine. When Mum had had enough of the water, she returned to sit with Dad. He had fallen asleep and was scarlet. She glanced around and realised her handbag was missing. It must have been stolen. Mum was furious with dad. Everyone hurried to the police station. Much to their surprise it had already been handed in and nothing was missing. The policeman said an old lady had found it in the beach toilets. Then mum remembered; she had left it there. Mum apologised to Dad and bought him a huge ice-cream.

Reading comprehension score = Total number of questions answered correctly

Reading summarisation score = Total number of main points recalled correctly

Phase 3 analysis

Same statistical methodology as Phase 2 of project:

- Raw scores were converted to age adjusted z- scores ($M=0$, $SD=1$) derived from normally hearing comparison group
- Regression analyses tested the effects of early confirmation on outcomes while controlling for effects of confounding variables
- Analyses run:
 - With and without BSL users (n=6)
 - With and without CI users (n=15)

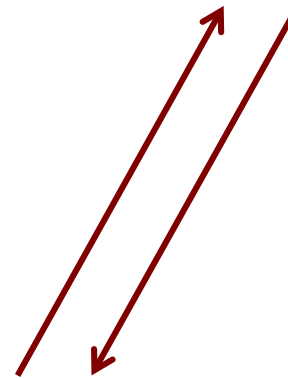
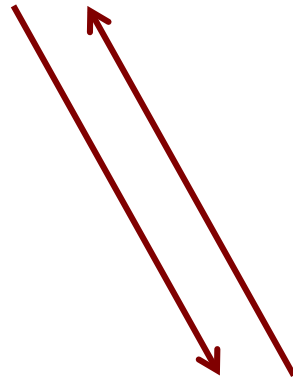
exposure



outcome

- a UNS programme
or
- confirmation by 9 months

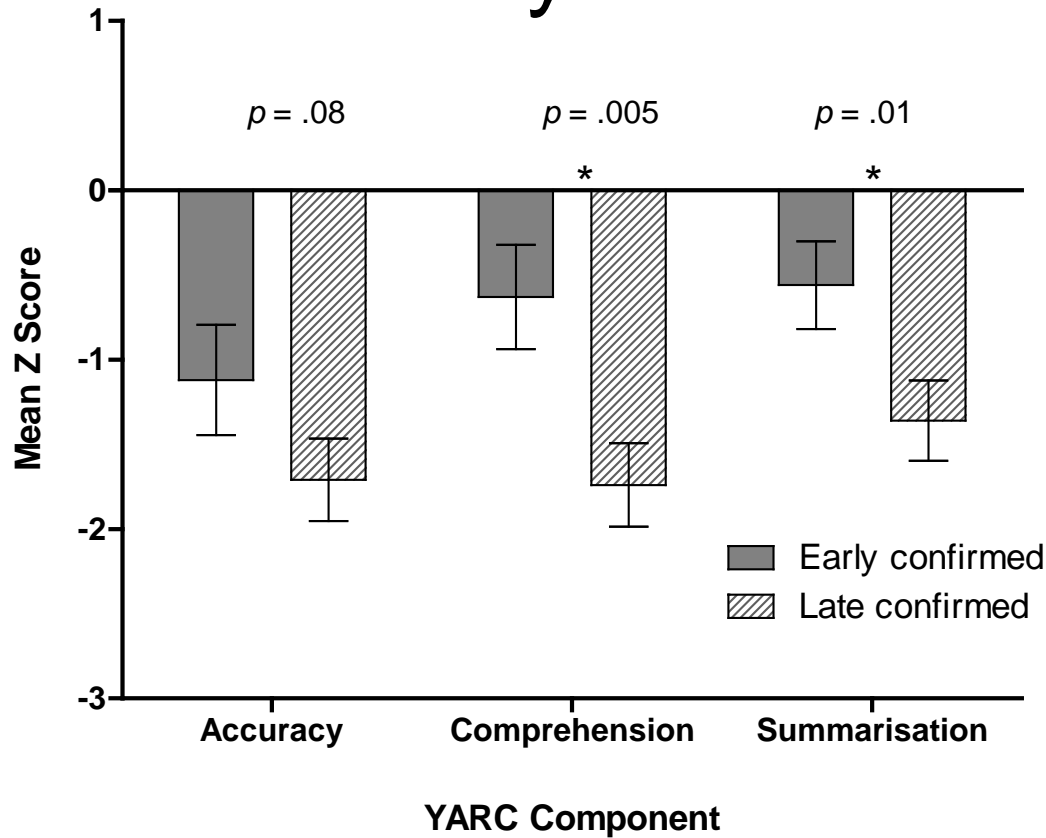
- 1° - READING COMPREHENSION
- 2° - speech, language
behaviour, ToM,
cost



confounders

child and
family factors

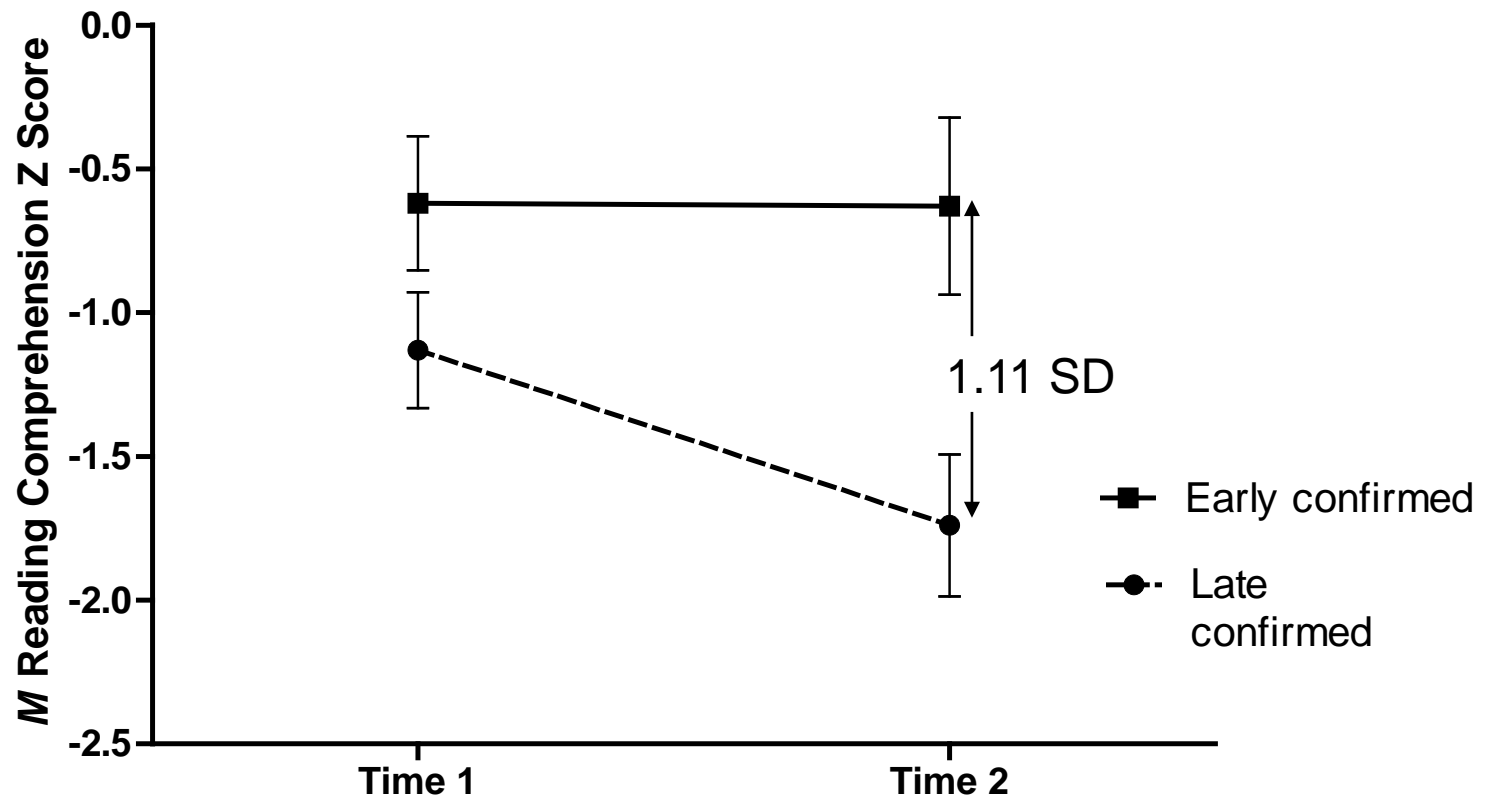
Phase 3 results: Reading early vs. late confirmed



* = significant adjusted mean difference in regression model that included

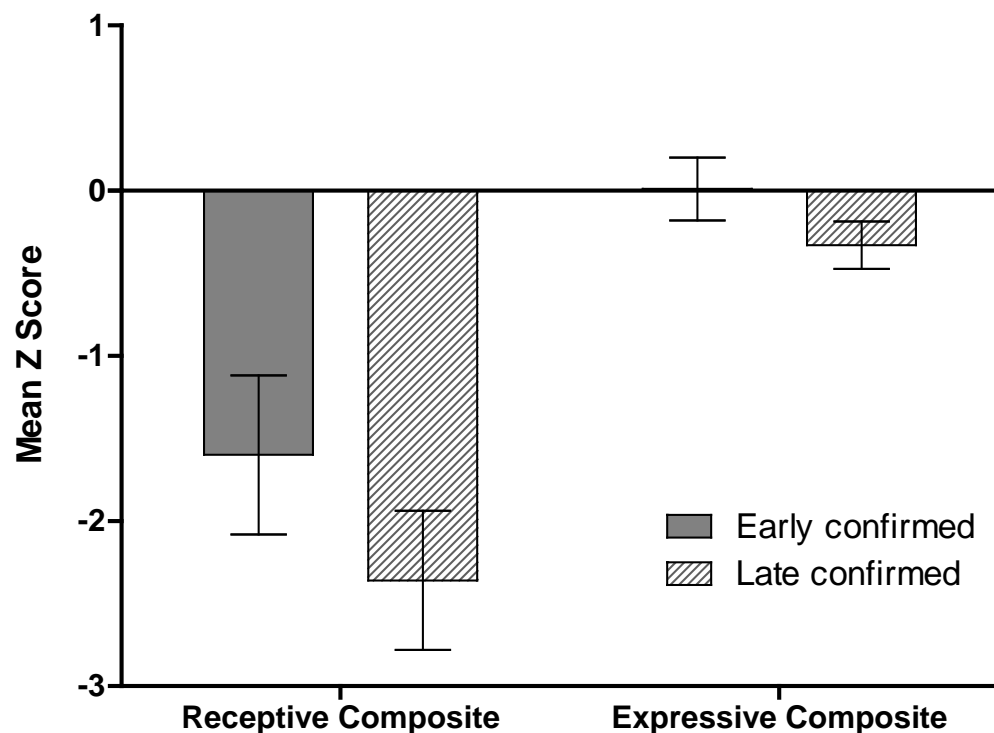
- non-verbal IQ
- maternal education
- severity of PCHI
- English as first language

Reading comprehension in phase 2 and phase 3



Mean unadjusted reading comprehension z scores at age 6-10 yrs (Time 1) and at age 13-19 yrs (Time 2) by age at confirmation of PCHI: confirmed early (< 9 months) versus confirmed late (>9 months)

Phase 3 results – language early vs. late confirmed



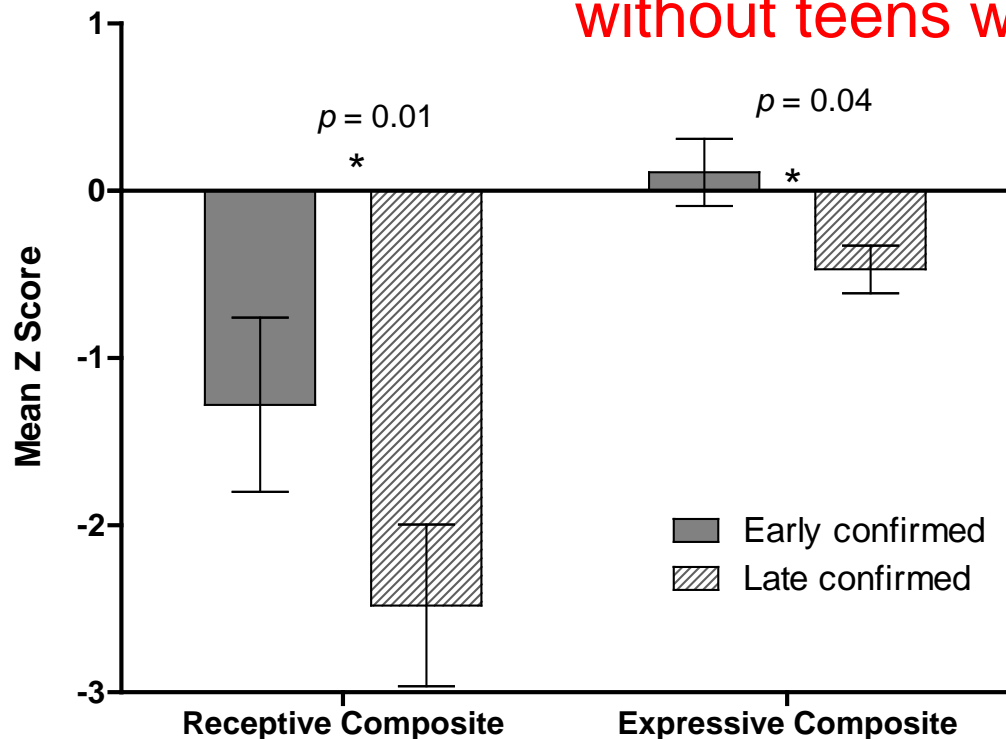
No significant adjusted mean difference in regression model that included

- non-verbal IQ
- maternal education
- severity of PCHI
- English as first language

Early confirmed $n = 27$
Late confirmed $n = 33$

Phase 3 results – language early vs. late confirmed.

without teens with CI



* = significant adjusted mean difference in regression model that included

- non-verbal IQ
- maternal education
- severity of PCHI
- English as first language

Early confirmed: $n = 21$

Late confirmed: $n = 27$

mean z scores and adjusted mean differences in children with early or later confirmation of PCHI: phase 2 results

| measure | n per group | mean (SD) of z score confirmation | | adjusted* mean diffs and 95% CI | P |
|------------------------------|-------------|-----------------------------------|-------------|---------------------------------|-------|
| | | by 9 mo | > 9 mo | | |
| speech | 45,52 | -1.24 (1.5) | -1.38 (1.6) | 0.29 (-0.28 to 0.87) | 0.32 |
| receptive language | 46,58 | -1.76 (1.5) | -2.37 (1.7) | 0.76 (0.26 to 1.27) | 0.004 |
| receptive/Ravens PCM diffs | 45,57 | -0.82 (1.2) | -1.66 (1.4) | 0.82 (0.3 to 1.3) | 0.002 |
| expressive language | 39,49 | -0.59 (1.3) | -1.07 (1.2) | 0.50 (0.00 to 1.01) | 0.05 |
| expressive/ Ravens PCM diffs | 39,49 | 0.14 (1.3) | -0.50 (1.3) | 0.70 (0.1 to 1.3) | 0.02 |

*adjusted for severity of hearing loss, maternal educational qualifications +/- Ravens PCM

in what way does early treatment improve expressive language? phase 2 findings.

| Outcome measure | Adjusted mean difference, model <u>not</u> including age aided (95% CI) | p | Adjusted mean difference, model including age aided (95% CI) | p |
|--|---|-------|--|--------|
| No. of sentences | 2.86 (0.49, 5.24) | 0.019 | 3.69 (1.04, 6.34) | 0.007 |
| Morphological endings | | | | |
| all | 0.68 (-0.04, 1.40) | 0.065 | 1.20 (0.20, 1.83) | 0.015 |
| low frequency | 0.19 (-0.86, 1.24) | 0.714 | 0.46 (-0.74, 1.66) | 0.451 |
| high frequency | 6.64 (1.96, 11.31) | 0.006 | 9.56 (4.50, 14.62) | <0.001 |
| | Adjusted odds ratio, model <u>not</u> including age aided (95% CI) | p | Adjusted odds ratio, model including age aided (95% CI) | p |
| No. of sentences with multiple clauses | 1.57 (0.67, 3.71) | 0.302 | 2.47 (0.89, 6.80) | 0.026 |
| Use of phonological simplifications | 0.56 (0.20, 1.61) | 0.283 | 0.23 (0.06, 0.84) | 0.026 |
| Narrative structure | 3.03 (1.09, 8.46) | 0.034 | 3.96 (1.21, 12.93) | 0.023 |
| Narrative content | 4.43 (1.52, 12.89) | 0.006 | 9.68 (2.60, 36.07) | 0.001 |

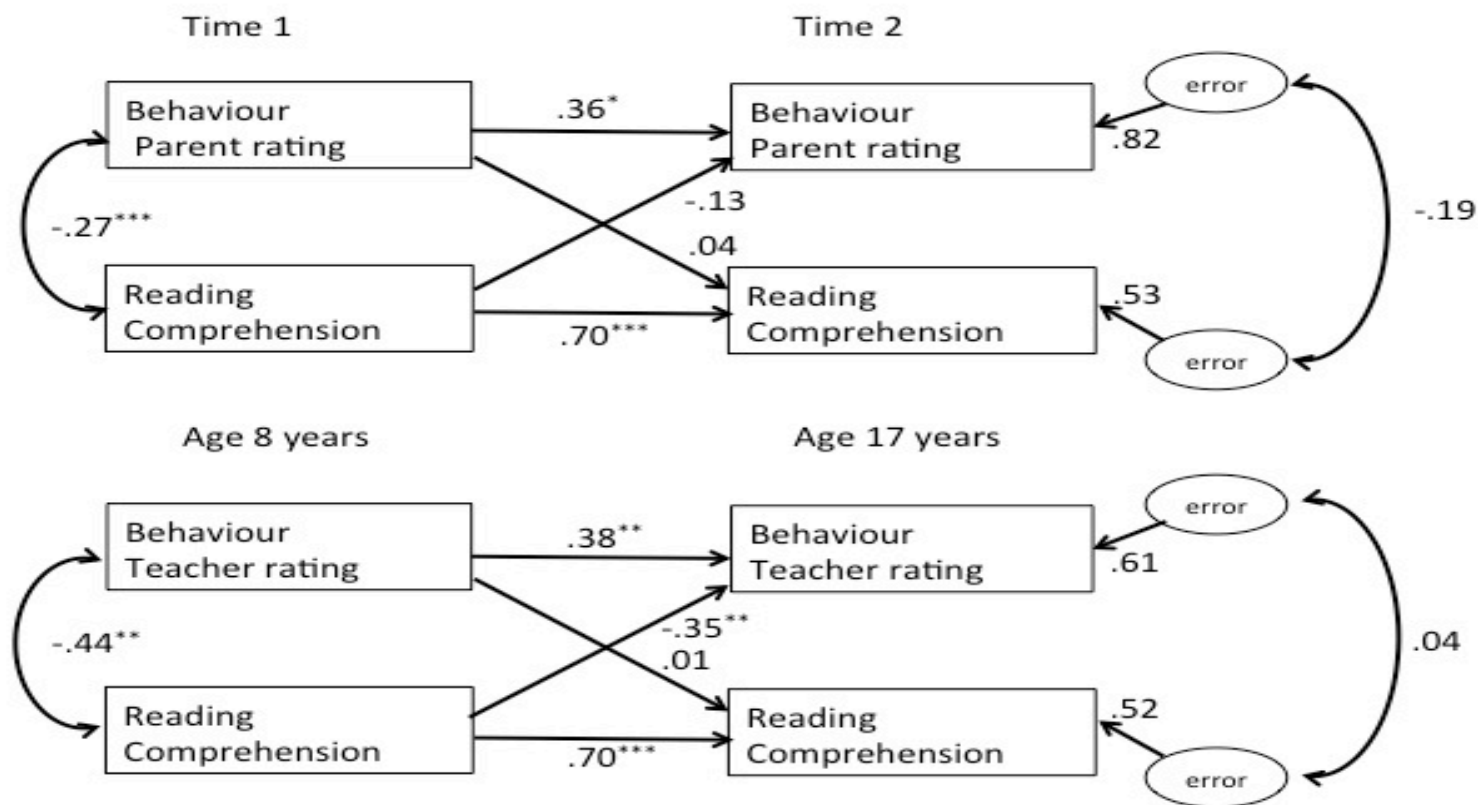
Mean Reading Comprehension, Language Comprehension, and EBD scores of children with PCHI at Time 1 (6-10 yrs) and Time 2 (13-20 yrs)

| Outcome Measure | n | T1 Mean | T2 Mean | SMD T2-T1 (95% CI) | p |
|--------------------------------------|----------|--------------------|--------------------|---------------------------|----------|
| Reading Comprehension | 62 | -1.03 | -1.28 | -0.17 (-0.52 to 0.18) | .10 |
| Language Comprehension | 62 | -2.32 | -2.51 | -0.07 (-0.42 to 0.28) | .50 |
| Parent rated SDQ Total Difficulties | 72 | 9.22 | 8.29 | -0.16 (-0.48 to 0.17) | .18 |
| Teacher rated SDQ Total Difficulties | 53 | 7.75 | 6.21 | -0.28 (-0.48 to 0.17) | .06 |

Forced entry stepwise regression predicting reading comprehension scores at Time 2 for PCHI (n=53)

| | R ² | R ² Change | F | d.f | p |
|--|----------------|--------------------------|-------|------|-------|
| Step 1 | .43 | .43 | 37.91 | 1,51 | <.001 |
| Time 1 Reading comprehension | | | | | |
| Step 2 | .46 | .03 | 0.83 | 4,47 | .51 |
| English first language, Mother's education, Severity of hearing loss, Time 1 Non-verbal IQ | | | | | |
| Step 3 | .63 | .17 | 20.42 | 1,46 | <.001 |
| Time 1 Language aggregate | | | | | |

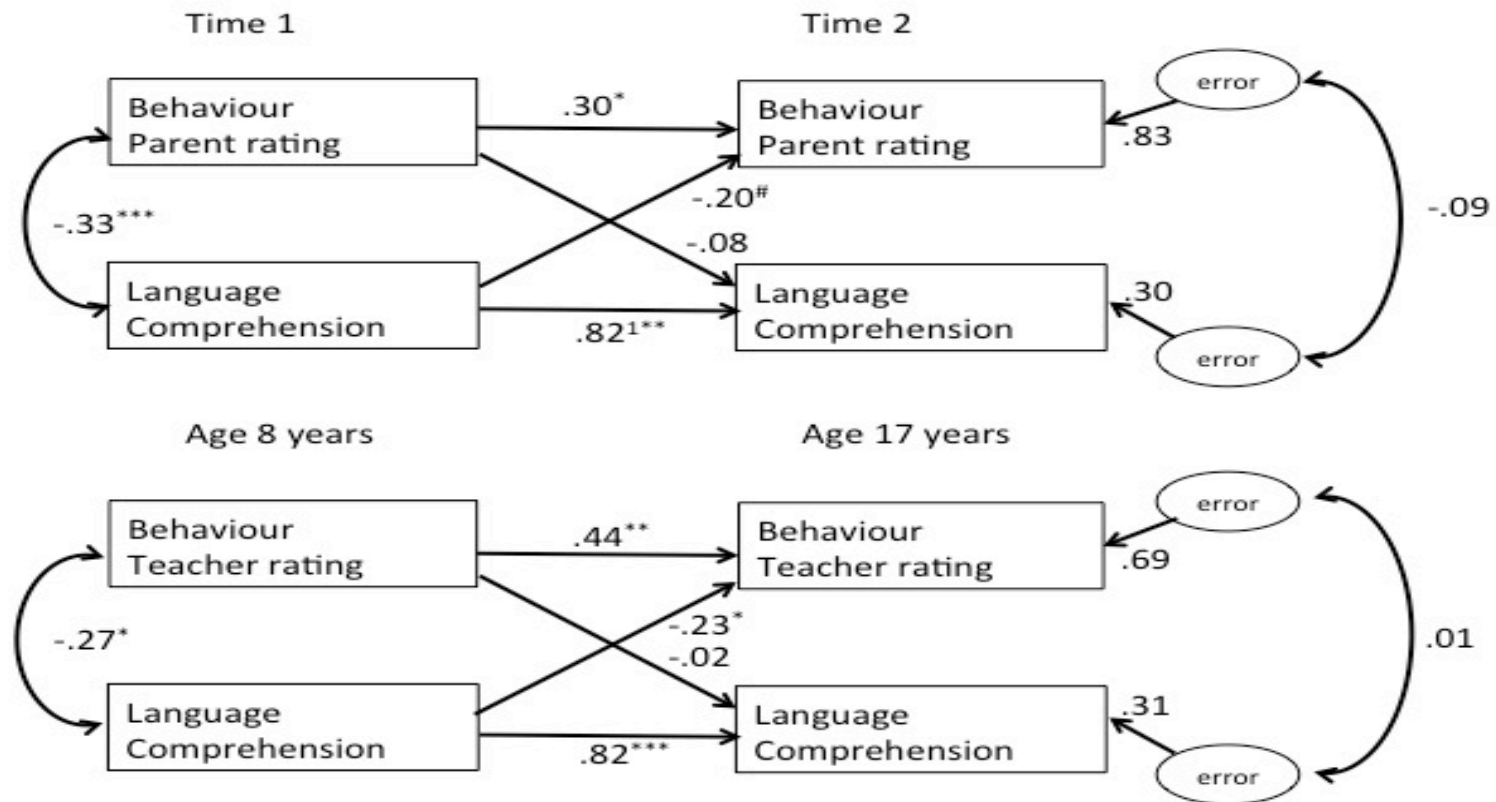
Parent and Teacher ratings of Total Difficulties on the SDQ and Reading Comprehension abilities for children with PCHI (n=62)



*** $p < .001$, ** $p < .01$, * $p < .05$

Standardised maximum likelihood estimates of cross-lagged models

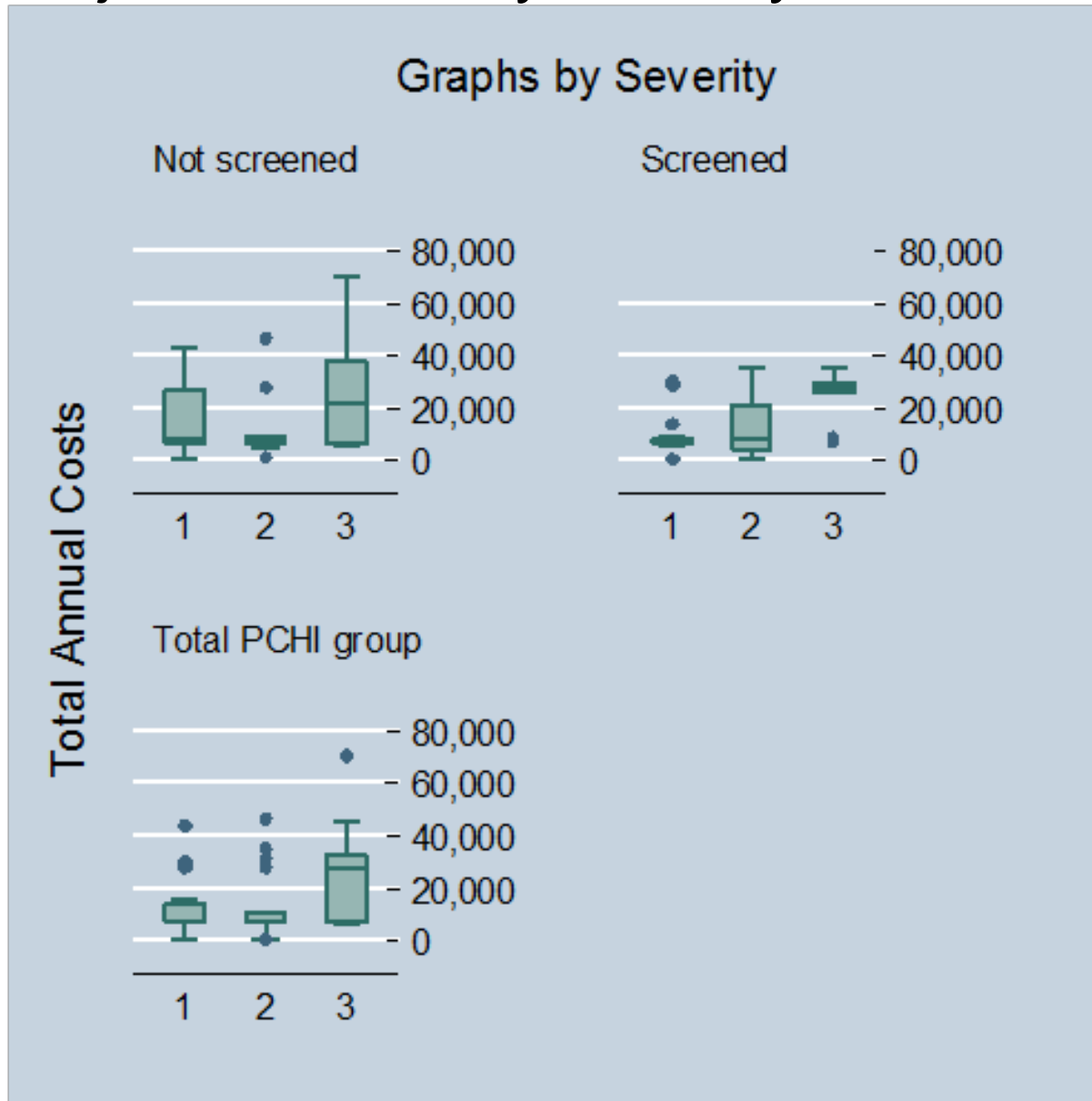
Parent and Teacher ratings of Total Difficulties on the SDQ and Language Comprehension in children with PCHI (n=62) at two time points.



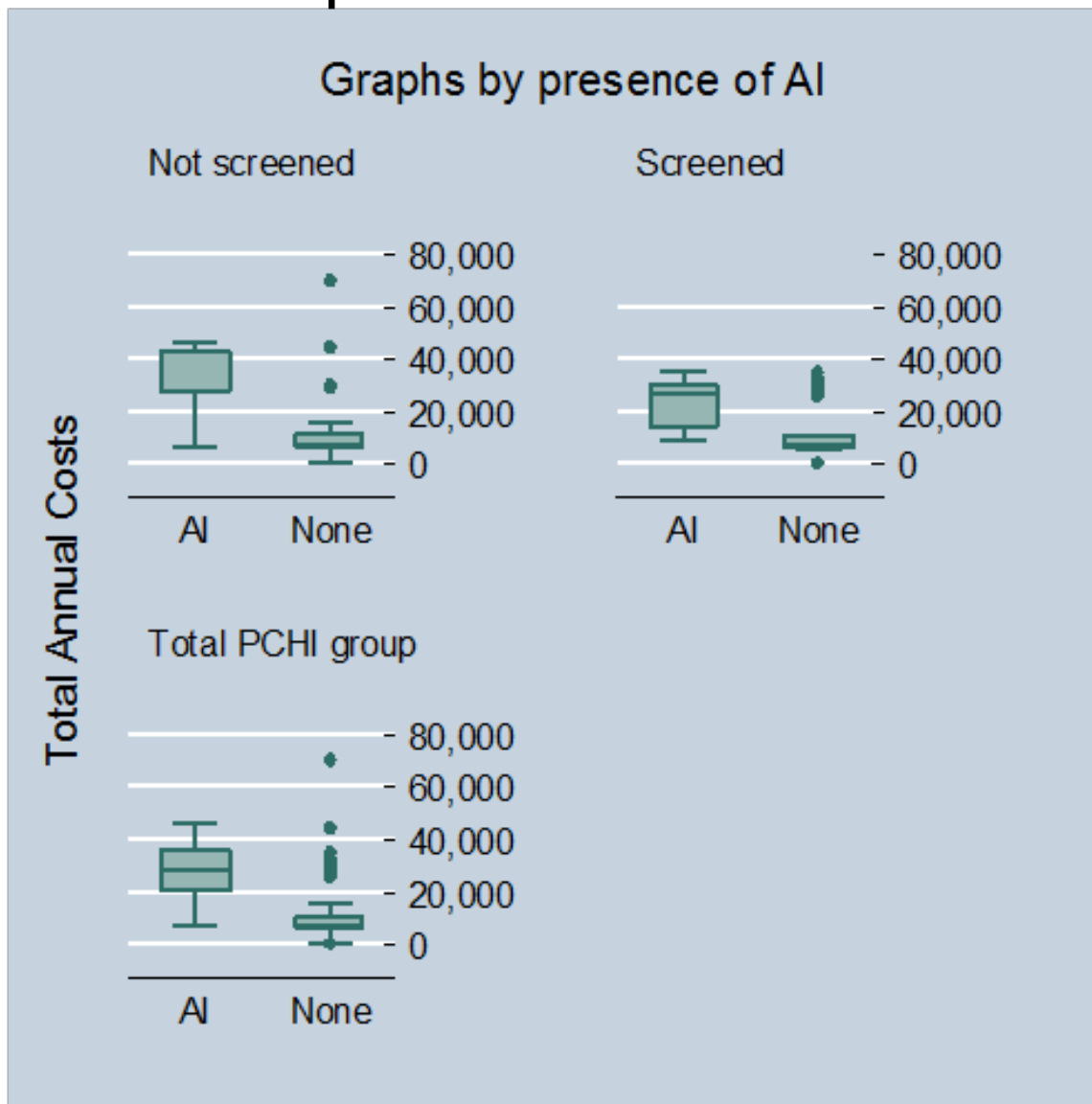
*** $p < .001$, ** $p < .01$, * $p < .05$, # $p < .08$

Standardised maximum likelihood estimates of cross-lagged models

Total unadjusted costs by severity: UNS and no UNS



Total unadjusted costs by presence of additional impairments: UNS and no UNS



strengths and weaknesses

strengths

- population based (95% of total births in 8 districts eligible)
- prospective and 'longitudinal'
- generalisable (8 districts)
- high rate of ascertainment
- multiple informants
- validated outcome measures
- own normative group
- assessment at home and blind to early history
- simultaneous adjustment for other explanatory factors

weaknesses

- not powered for subgroups
- variability of intervention (7 services); 1990s standards
- ? insensitive expressive language measures

Summary

Results from this analysis suggest:

1. Early confirmation of PCHI is associated with better reading and language comprehension in the teenage years.
2. The difference in reading comprehension skills between early- and late-confirmed deaf teenagers has widened since childhood.
3. The benefit of early confirmation on language outcomes is not apparent for teenagers who have received CI.
4. After taking childhood reading comprehension into account, childhood language still predicts teen age reading comprehension.
5. Childhood reading & language comprehension predict teenage emotional and behavioural disorders not vice- versa
6. There are trends towards reduction in societal costs after UNS

Thank you!

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wellcometrust

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