

Feeding Difficulties



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The RASopathy Specialist MDT

General Pediatrician

Complex Care CNS

Initial Consultation

Cardiology

Pediatrician

Neurodevelopment

Endocrine

Orthopedics

Genetics

Follow up every 3 years



Ang Roberts (Gen)



Juan Kaski (Cardio)



Anish Sanghrajka (Ortho)



Lee Hudson (Res)



Elaine Clark (Dev't)



Mehul Dattani (Endo)



Harshini Katugampola (Endo)



Keir Shiels (Lead)

Rasopathy Management

Medical
Background

Growth and
Puberty

Cardiac Status

Genetic
Assessment

Vision and
Hearing

Hair and Teeth

Skin

Musculoskeletal
Development

Bleeding Risk

Social and
Educational
Development

Feeding and
Nutrition

Clinical Priorities for Noonan Families are not reflected proportionately in the research literature

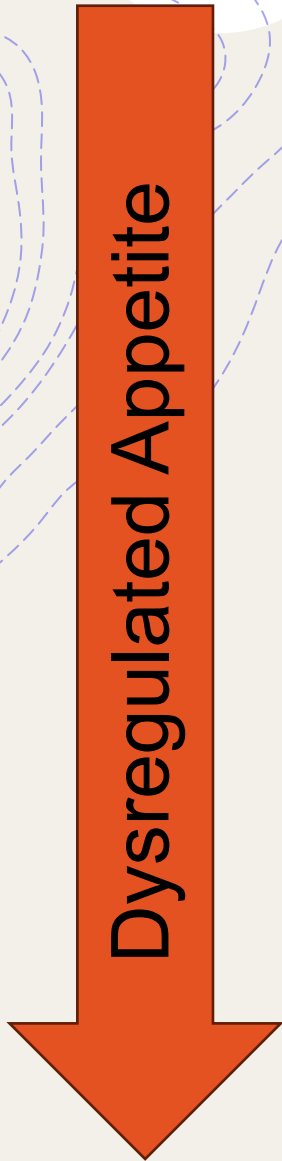
	Proportion of families declaring "Major problem impacting life"*	PubMed Results searching for "Noonan Syndrome + Problem"
 Cardiac	5.8%	1286
 Growth	19.2%	679
 Leg Pain†	15.4%	14
 Feeding Difficulties	26.9%	56

*Families were asked to grade symptoms based on daily impact to quality of life:
Major, Significant, Manageable, Occasional, Minimal

†In literature review, of 16 sources, all 16 mention Pulmonary stenosis as a feature of Noonan Syndrome, but only 6 mention extremity/joint pain despite similar prevalence (65%) [2,3,4]

Problem	Percentage reporting that healthcare professionals were dismissive of the problem
Leg pains	29.03%
Feeding difficulty	24.14%
Looking or feeling different from other children	22.58%
Behavioural difficulty	19.35%
Puberty	14.81%
Fine motor skills	12.90%
Learning difficulty	12.50%
Growth issues	4.76%
Cardiac issues	2.04%

Table 17: *The percentage of patients reporting that healthcare professionals were dismissive of the problem for each issue.*



Dysregulated Appetite

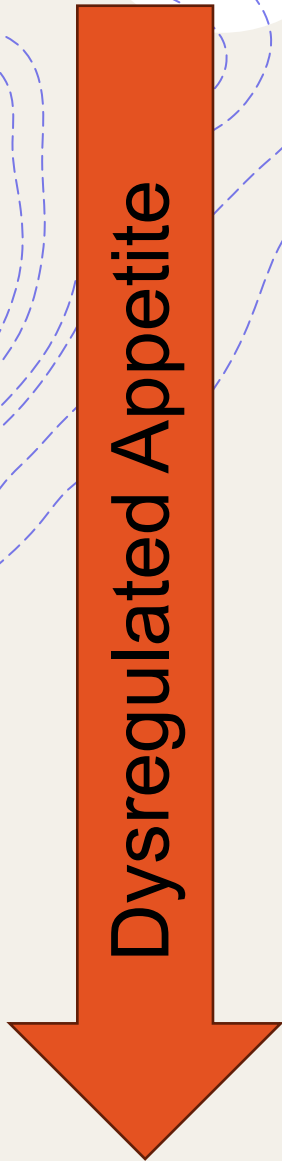
Birth – Cardiac, Chylothorax

0-2 years - Vomiting

1-4 years – Oral aversion

4 years on – Textural, Visual
& smell sensitivities

Dysregulated Appetite



Birth – Cardiac, Chylothorax

Diagnostic overlay &
Diagnostic overshadowing

0-2 years - Vomiting

Misdiagnosis and
mismanagement
(Reflux, milk allergy...)

1-4 years – Oral aversion

Feeding Adjuncts (NG, NJ,
PEG)

4 years on – Textural, Visual
& smell sensitivities

Autism Spectrum
ARFID
Fussy Eating

Feeding Problems



LOW APPETITE
'GRAZING'



LOW STOMACH
CAPACITY 'FULL
QUICKLY'



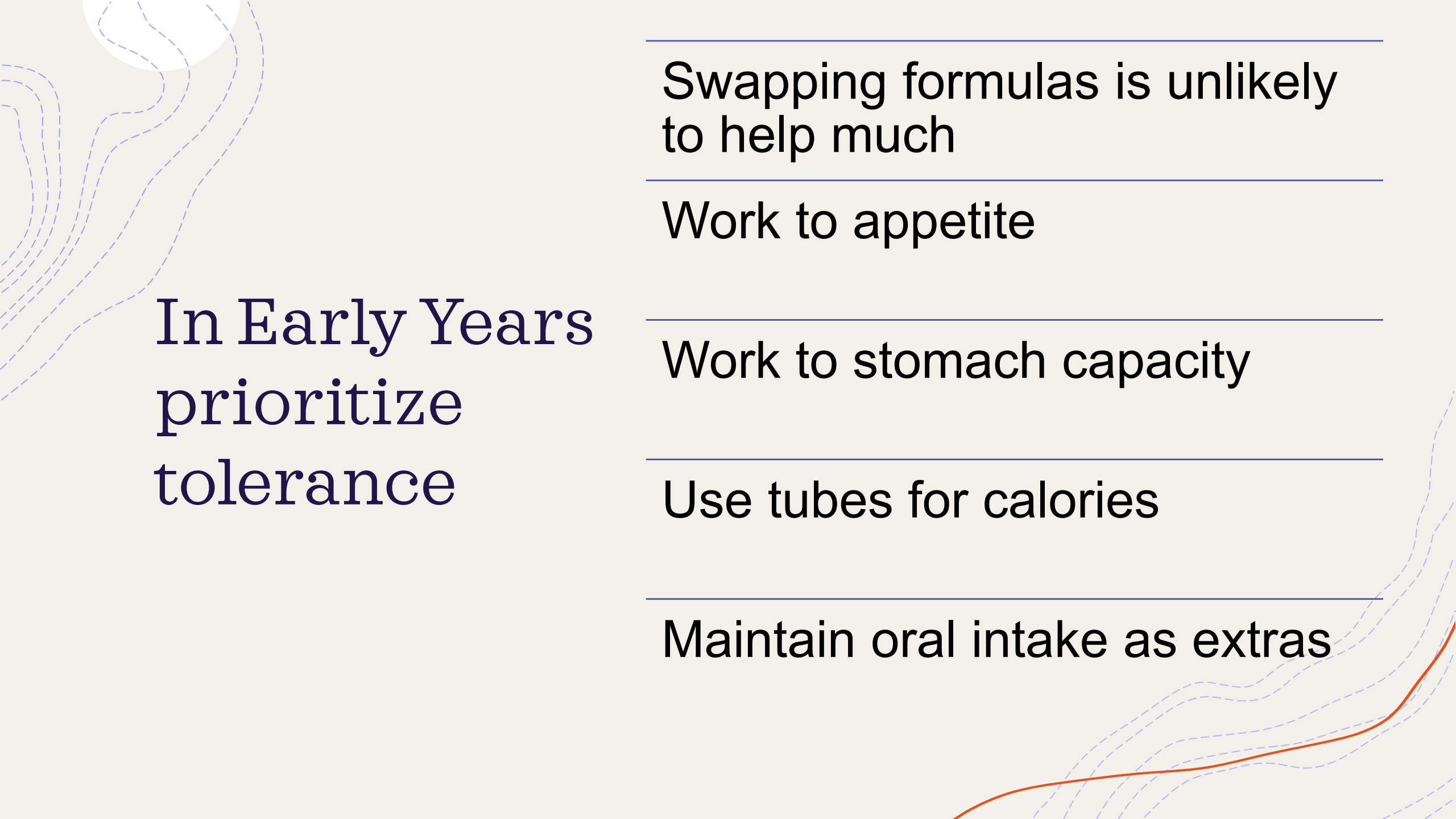
SLOW GUT
TRANSIT



QUICK
VOMITING



STEP BACK
WITH
ILLNESSES



In Early Years
prioritize
tolerance

Swapping formulas is unlikely
to help much

Work to appetite

Work to stomach capacity

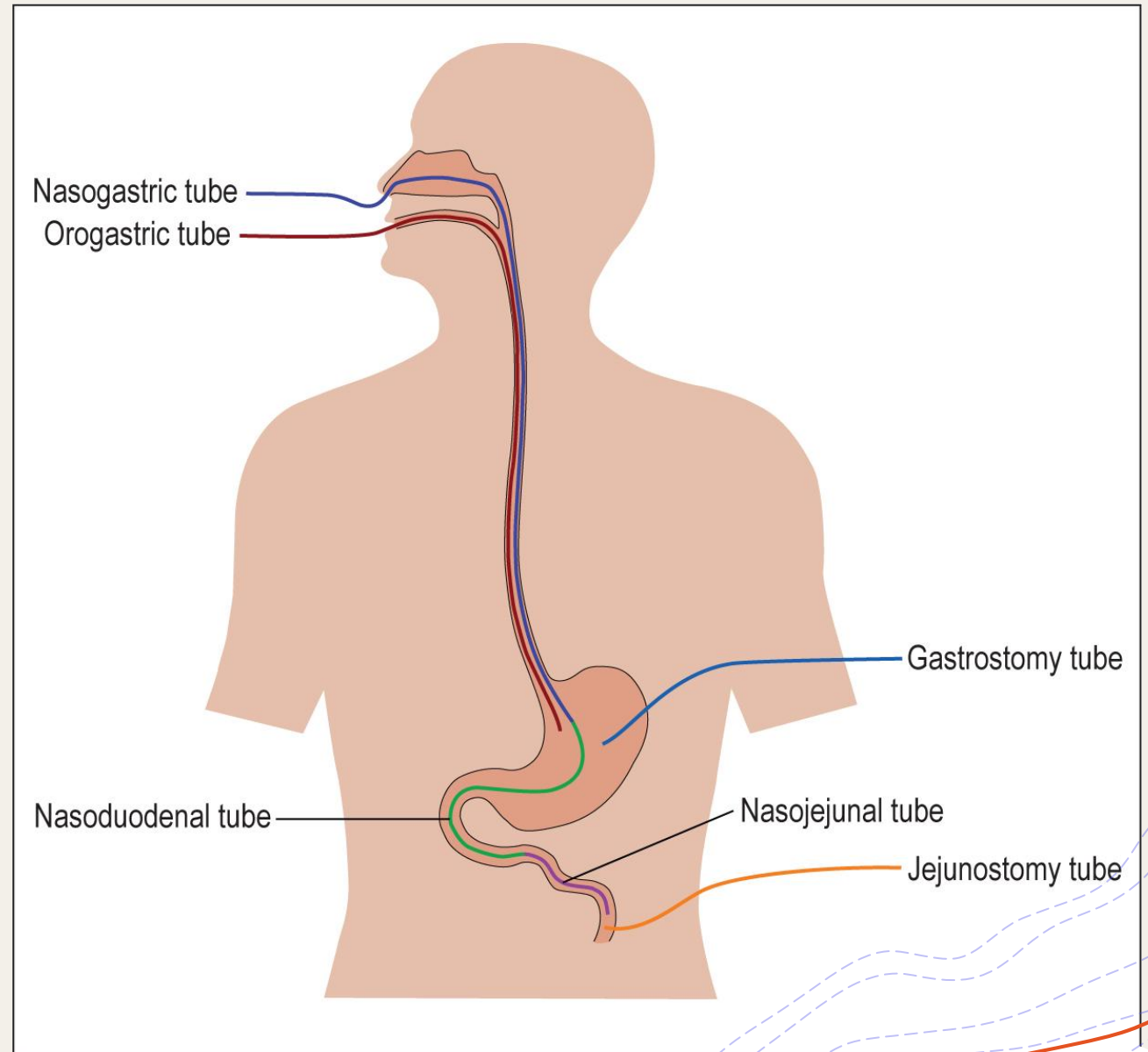
Use tubes for calories

Maintain oral intake as extras

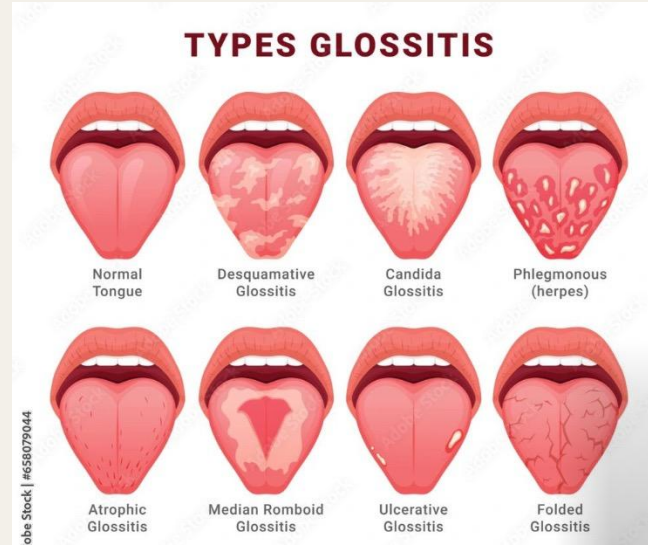
Cyproheptadine: Appetite stimulant

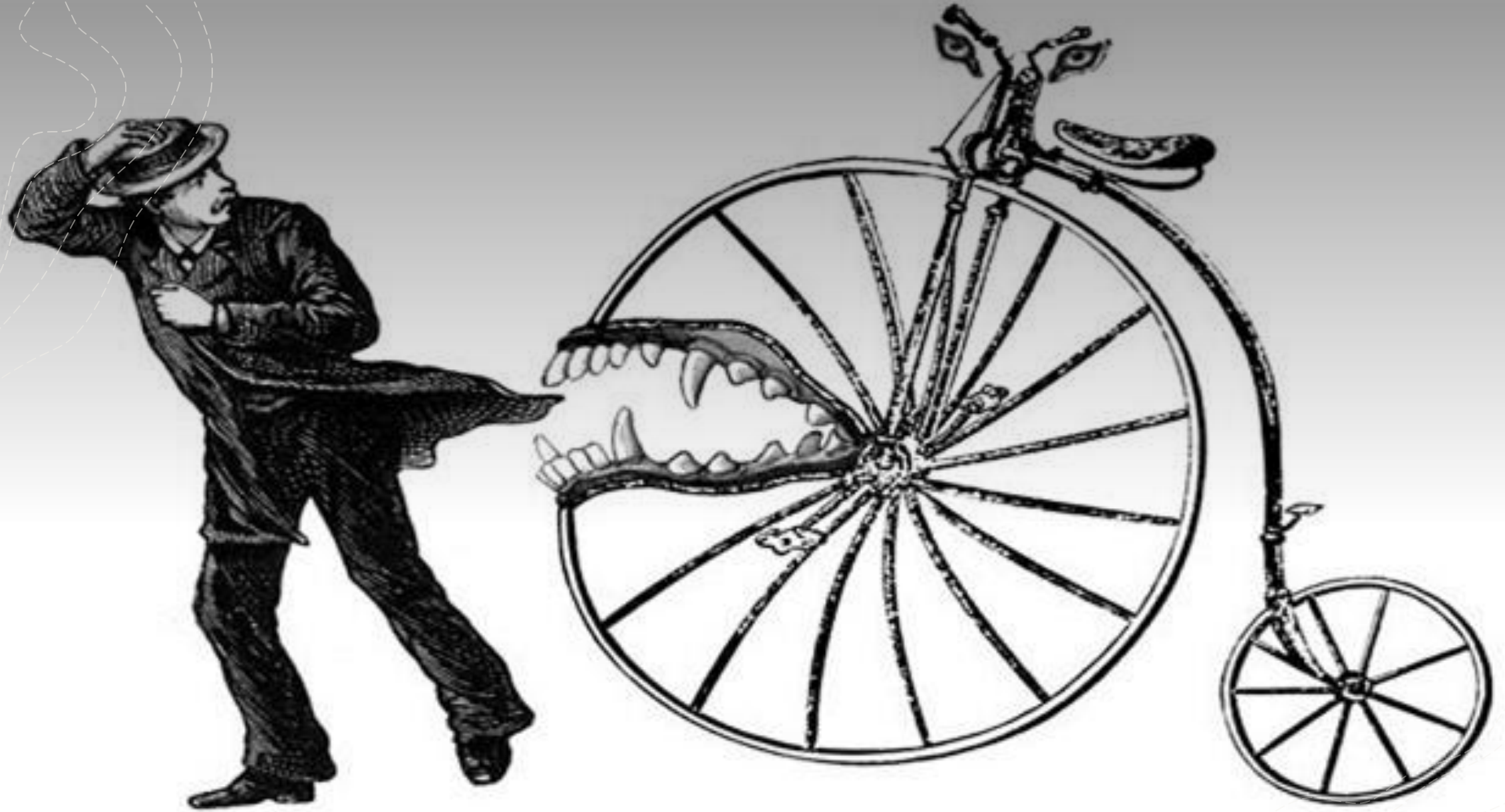


Types of tube



Effects of feeding difficulties





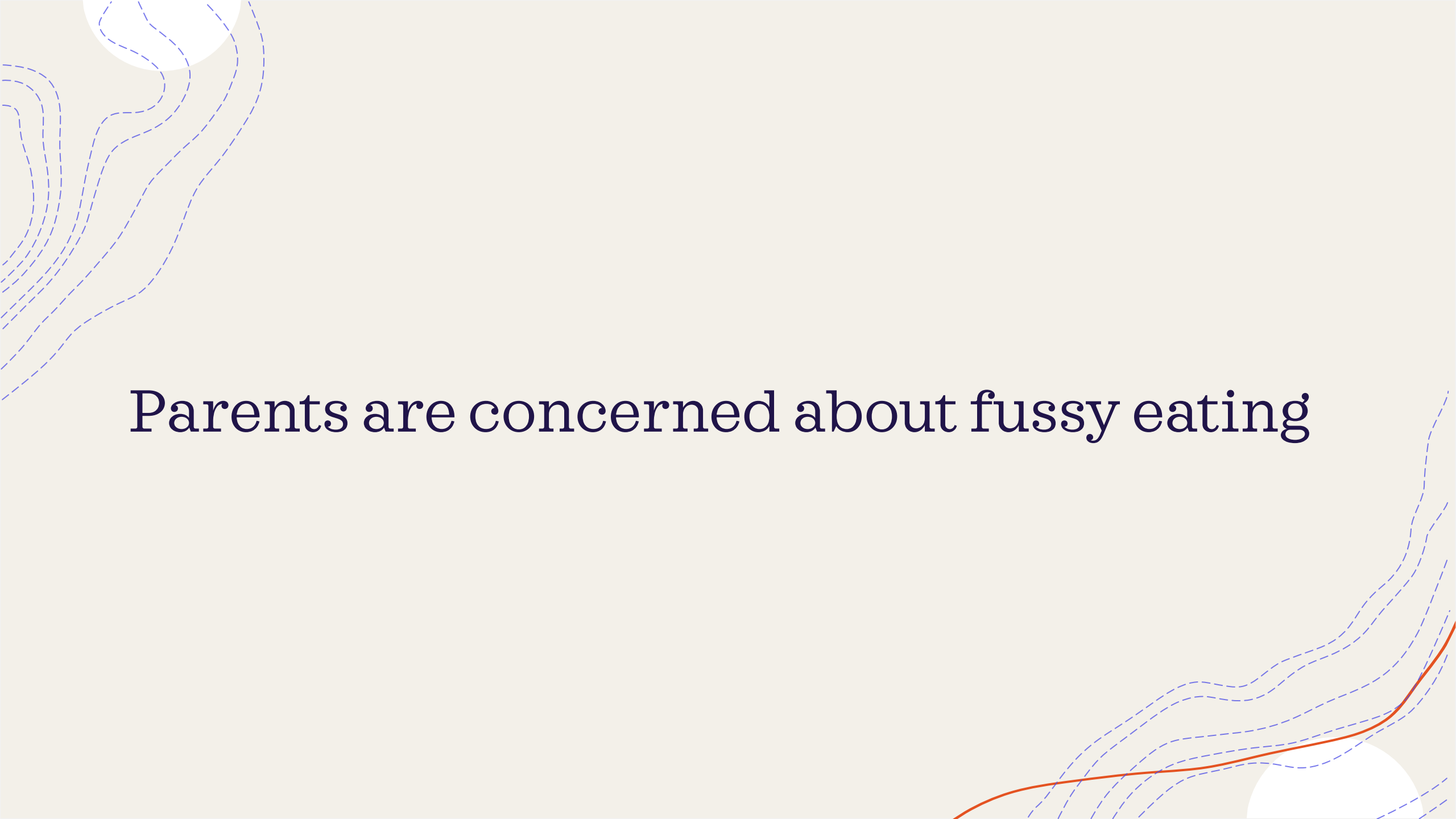
A VICIOUS CYCLE

Repeating
unpleasant
things creates
fear

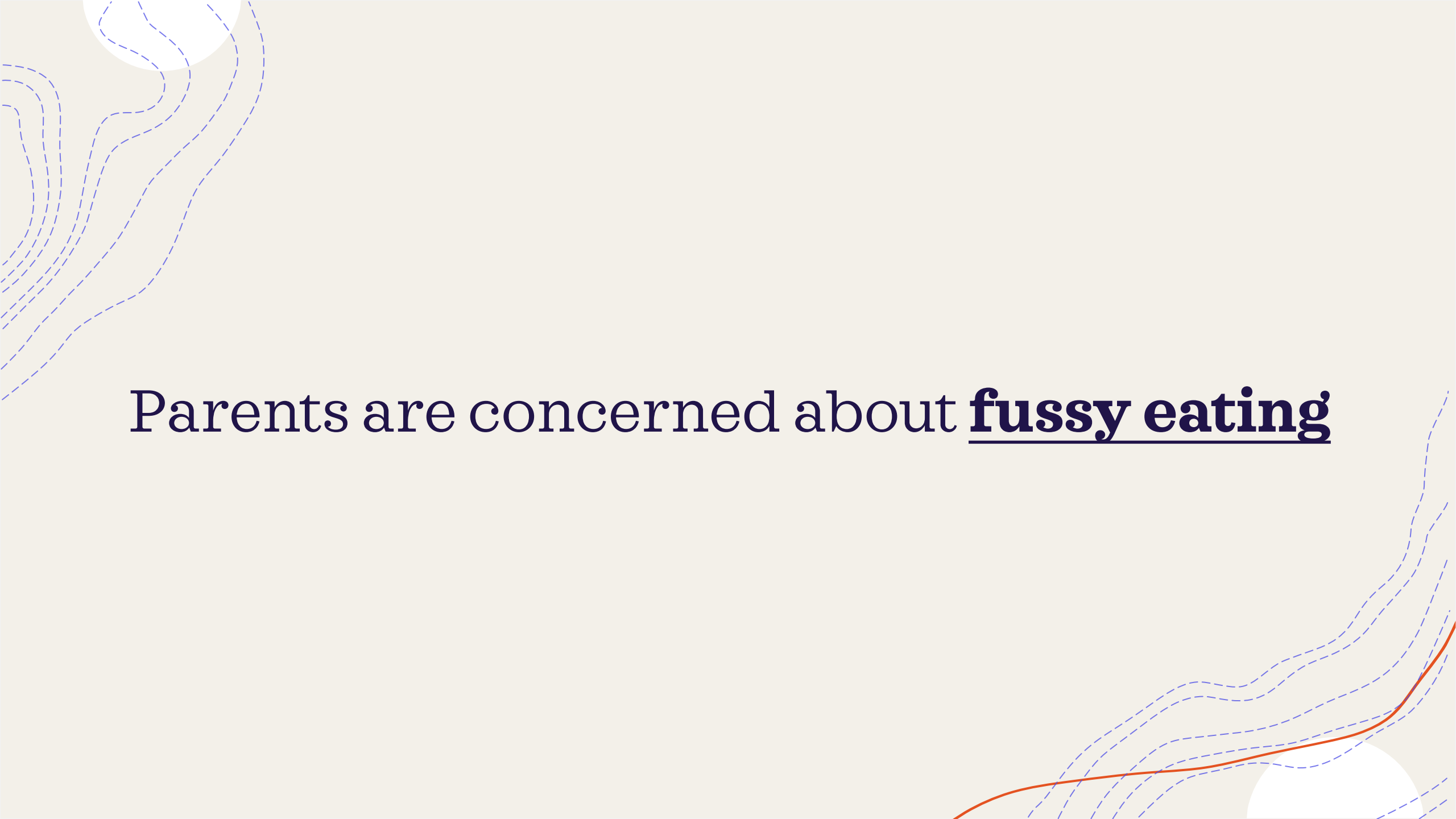


FEAR

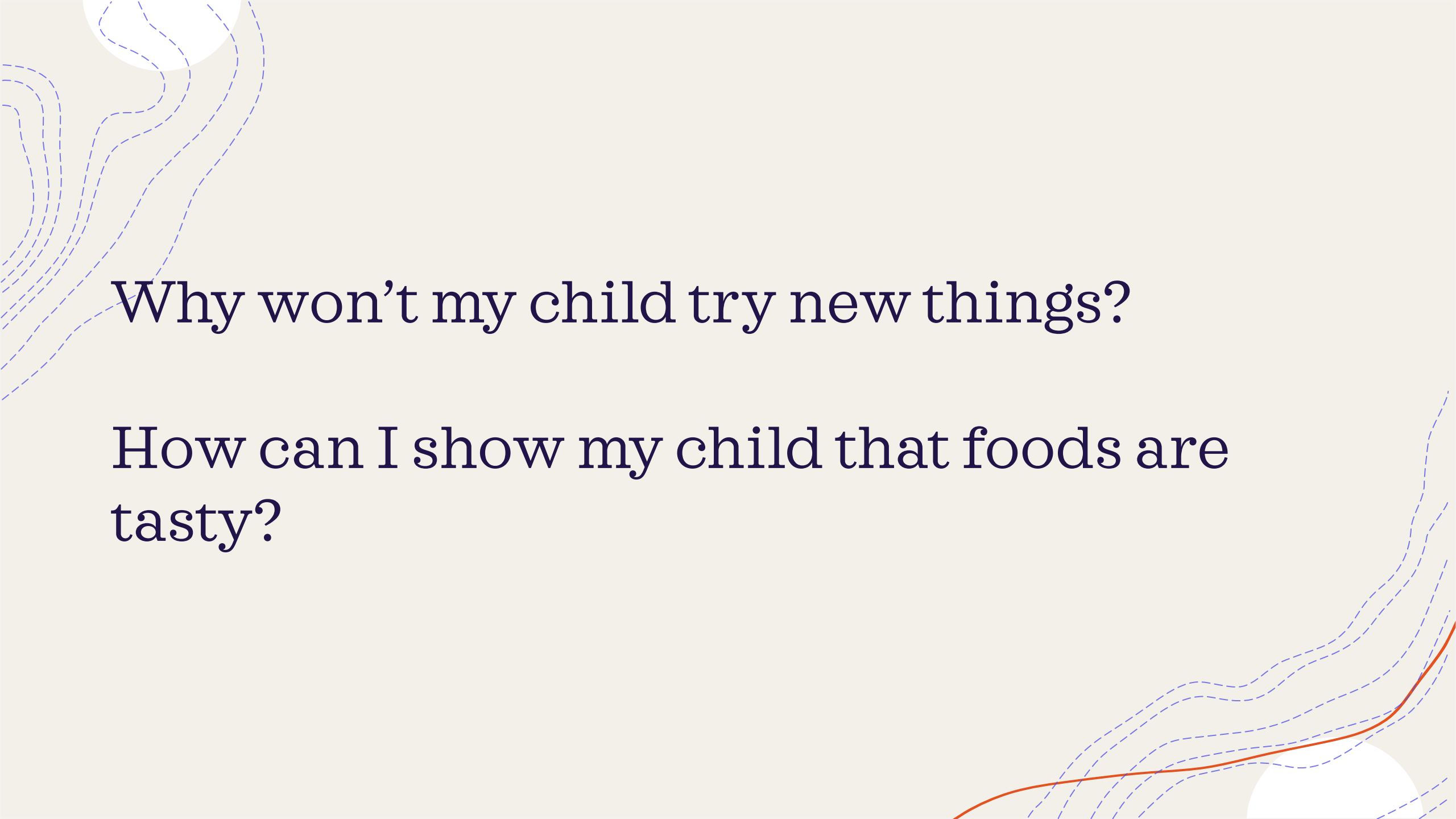
#INSIDEOUT

The background is a light beige color. In the top-left corner, there is a white circle partially cut off by the edge, with several blue dashed wavy lines flowing downwards and to the right from it. In the bottom-right corner, there is another white circle partially cut off, with several blue dashed wavy lines flowing upwards and to the left from it. A solid orange line also flows from the bottom-right towards the center, following the general direction of the blue lines.

Parents are concerned about fussy eating

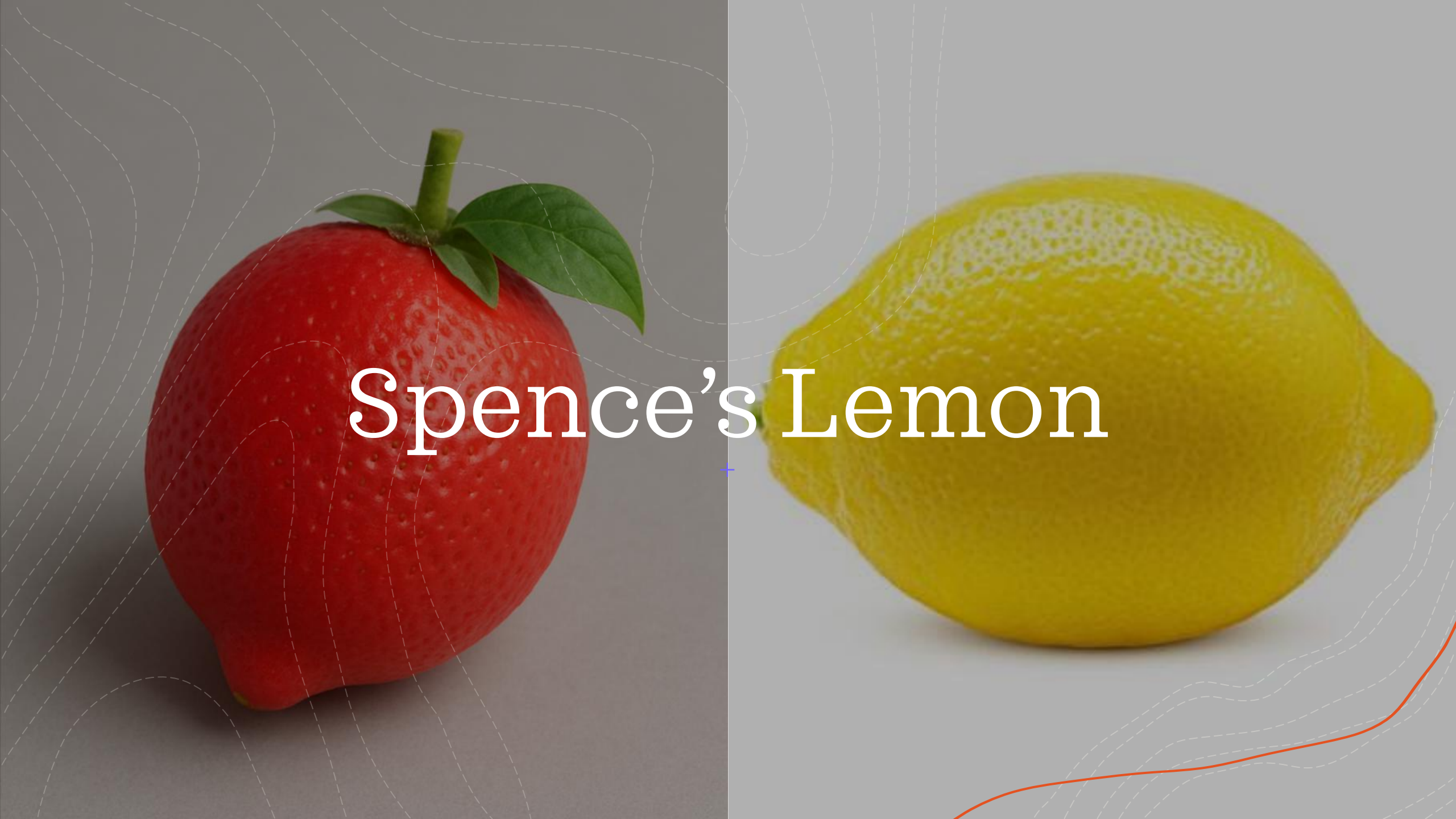
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Parents are concerned about **fussy eating**

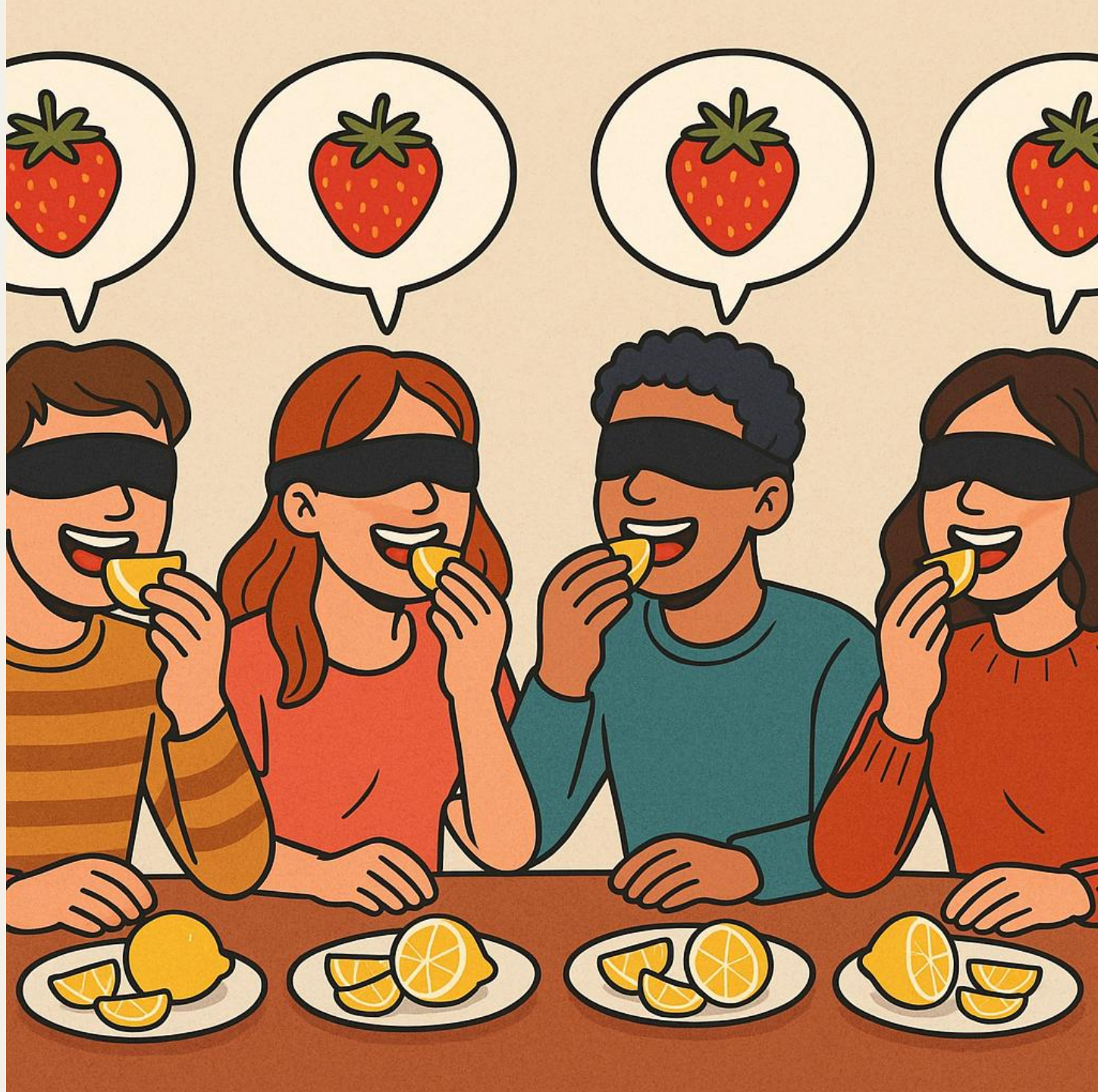
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Why won't my child try new things?

How can I show my child that foods are
tasty?



Spence's Lemon



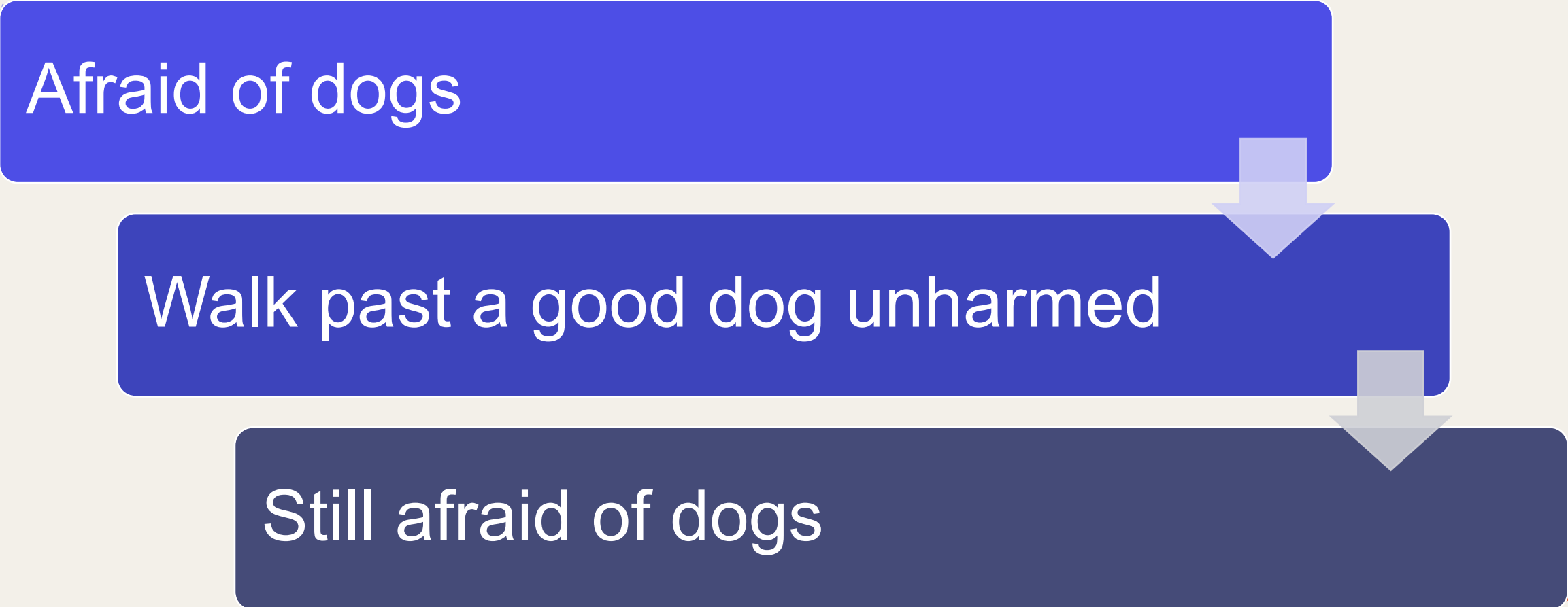




Expectation Alters Perception

Exposure to a pleasant surprise is
not aversion therapy

Afraid of dogs



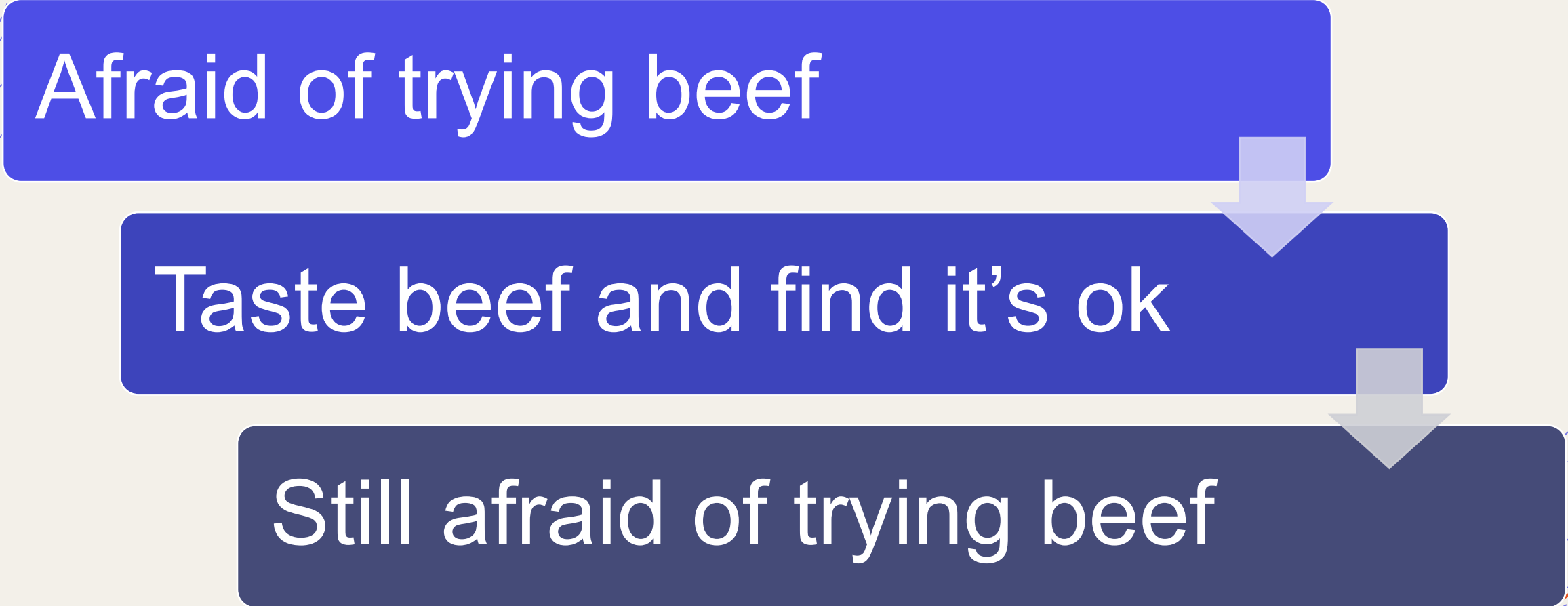
```
graph TD; A[Afraid of dogs] --> B[Walk past a good dog unharmed]; B --> C[Still afraid of dogs];
```

Walk past a good dog unharmed

Still afraid of dogs

Exposure to a pleasant surprise is
not aversion therapy

Afraid of trying beef



```
graph TD; A[Afraid of trying beef] --> B[Taste beef and find it's ok]; B --> C[Still afraid of trying beef];
```

Taste beef and find it's ok

Still afraid of trying beef



Perception does
not easily alter
expectation

The background features a light beige color with several thin, wavy, dashed lines in a muted blue-grey tone. There are also two solid white circles: one in the top-left corner and another in the bottom-right corner. A single solid orange line curves along the bottom right edge of the frame.

Coercion is
Counterproductive

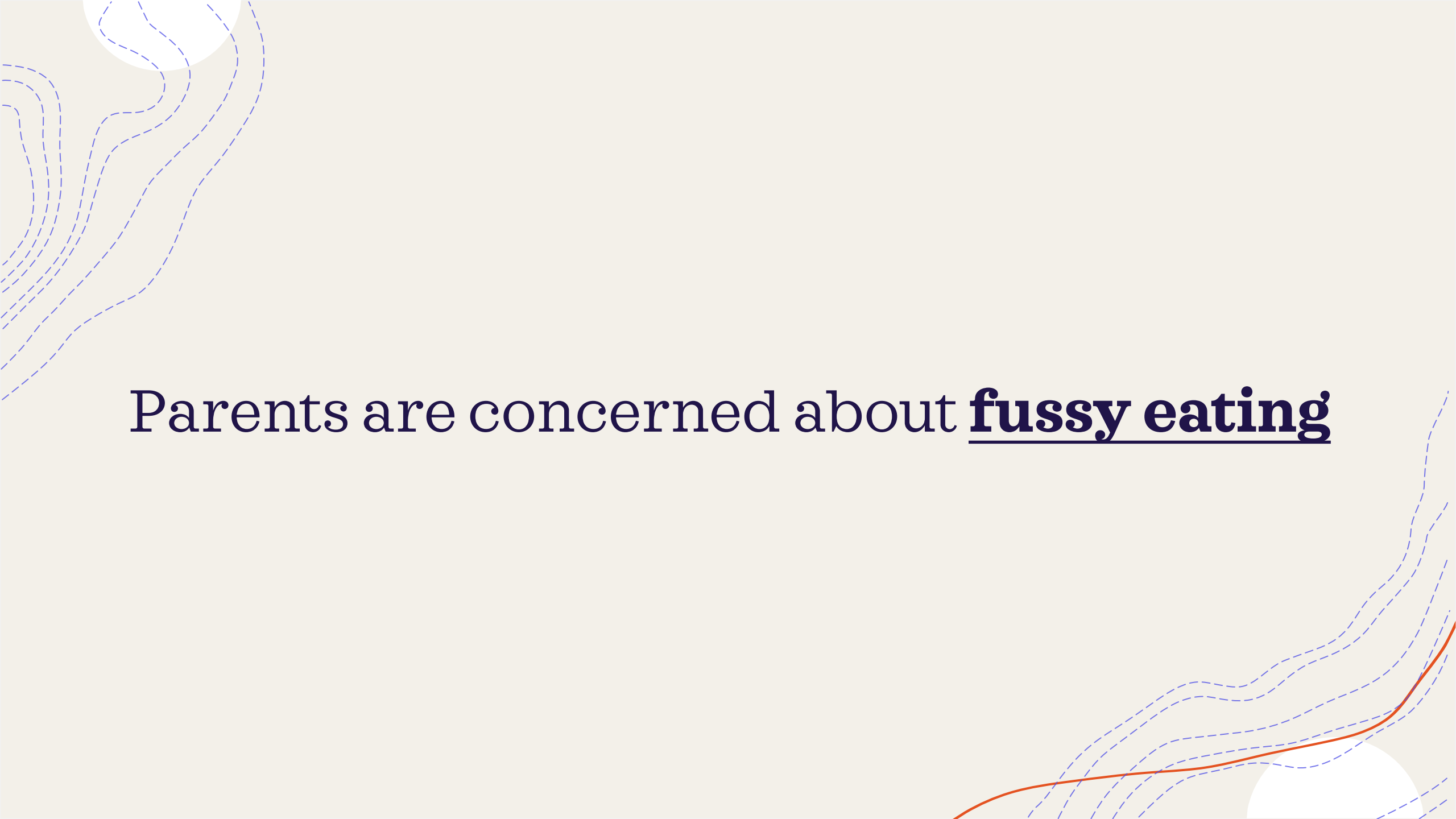
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Why are you worried? Really?

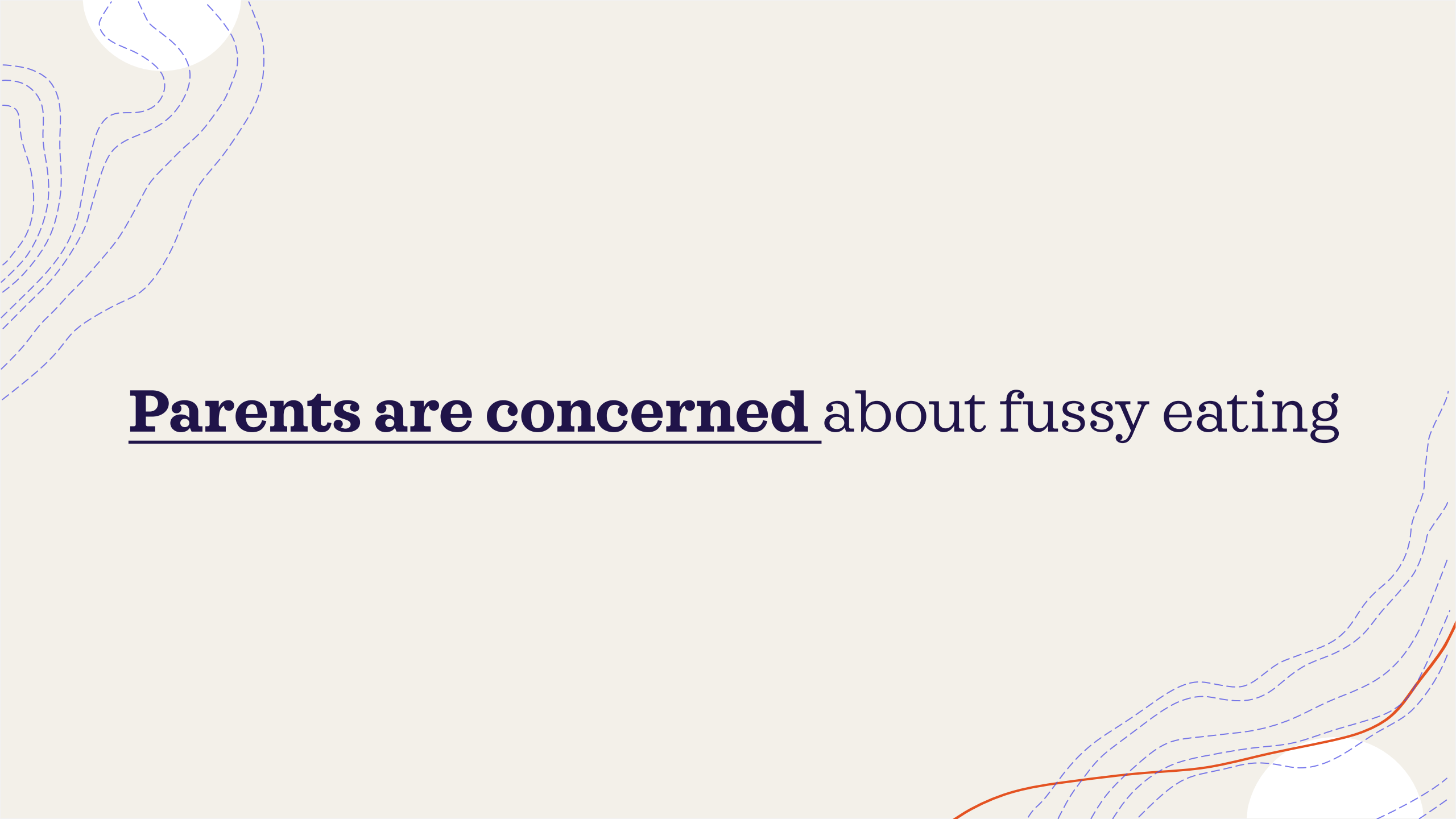




Agitation Alters Perception

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Parents are concerned about **fussy eating**

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Parents are concerned about fussy eating

Professionals Referral Clinic

Psychologist

Psychiatrist

Paediatrician

Local
Dietician

Local
Paediatrician

Local OT

Feeding and Eating Disorder Service

ARFID and Autism Spectrum Behaviours

Nutritional Management

Family Pressures

Reducing the child's stress around food

Expectation Management

Integrating plans with family, school and local services



Outcomes

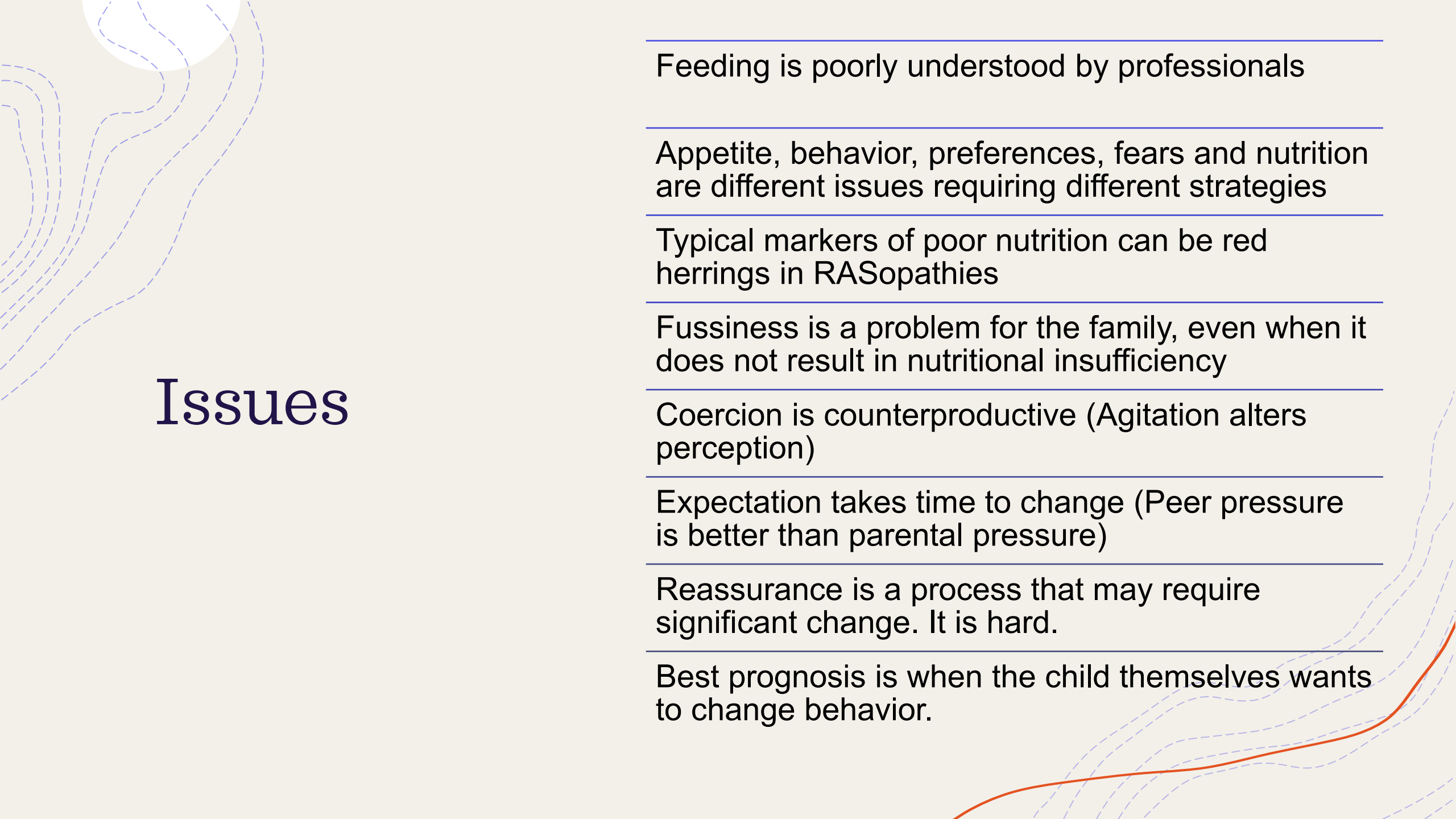
Autism Assessments

Family Therapy

Feeding and Eating Disorders
Service

Advice and plan

Reassurance



Issues

Feeding is poorly understood by professionals

Appetite, behavior, preferences, fears and nutrition are different issues requiring different strategies

Typical markers of poor nutrition can be red herrings in RASopathies

Fussiness is a problem for the family, even when it does not result in nutritional insufficiency

Coercion is counterproductive (Agitation alters perception)

Expectation takes time to change (Peer pressure is better than parental pressure)

Reassurance is a process that may require significant change. It is hard.

Best prognosis is when the child themselves wants to change behavior.