Health Care for All Persons with Down Syndrome

Persons with Down syndrome are living longer
- Recognition of specific health conditions, treatment and prevention
- Health promotion to physicians, families & individuals
- Increased availability & access to health care
- Improved opportunities for participation in clinical research

Improvements in health care have been “uneven”
- Limited by access to medical knowledge, resources or implementation of best practices
- Challenge of providing care to complex individuals
- Much to be done!
Need for Regular & Sustained Health Care

Increased prevalence of specific medical conditions
- Birth anomalies and medical conditions in newborn
- Acquired medical conditions in children & adolescents
- Acquired & age-related medical conditions in adults

Possible Barriers
- Down syndrome/ID - unfamiliar to primary care providers
- Best practices or Minimal standards? - what are they? How decided?
- Presentation - unusual, atypical, complicated picture
- Complexity - multiple, co-occurring conditions, overshadowing
Health Maintenance the Role of Screening?

Medical SCREENING of asymptomatic individuals

- Everyone with DS?
- Symptom based or high-risk individuals only

- Test BENEFITS, specificity, sensitivity, efficacy
- Potential COSTS, feasibility, harms or hassles
- Prevention of morbidity-mortality, improved QOL

Alternatives to SCREENING Everyone w/ DS

- Remain vigilant, monitor closely, ask the right Qs
- Maintain high suspicion based on age, risk factors
Regular & Sustained Health Care

Can screening improve health outcomes and quality of life

Yes - To what degree?

Depends on condition and functional outcome being measured
- Heart & Gastrointestinal defects (newborn)
- Leukemia, Thyroid disease (newborn, childhood)
- Vision & Hearing impairment (life long risk)

Better evidence needed
- Sleep disturbance, sleep apnea (life-long risk)
- Neurobehavioral & Mental Health disorders (life-long risk)
- Dementia (elderly)
Health Care for Down syndrome

AAP Health Supervision Guidelines (2011)
http://pediatrics.aappublications.org/content/128/2/393.long   English & Spanish Versions

NIH Down Syndrome Consortium
http://downsyndrome.nih.gov/Pages/default.aspx

Resources

NLM Medline Plus
AAP Health Supervision for Children with Down Syndrome


Based on expert opinion supported by published evidence

Provides guidance to Primary Care Providers across various settings (U.S.)

**Emphasis:** Screening & early diagnosis for common conditions
Increased vigilance & monitoring in asymptomatic individuals
Guidance specific to DS/ID
Access to best information and community resources

**Explicit Action items:** Assess, Evaluate, Measure, Administer, or Refer for Treatment
Infant & Young Child (1mo-5yr)

Actionable items based on physical findings and increased prevalence. No single child has all these conditions

Newborn Medical Follow-up: Cardiac repair (septum and valves), GI anomalies, Feeding, Respiratory, Vision, Hearing, Blood count, Thyroid

Ongoing Evaluations: Hearing, Vision, Thyroid (TSH/fT4), Iron (Ferritin), Growth rate, Dental, Development-Behavior, Sleep history - Vaccinations, Routine care

Sustained Vigilance: Feeding problems, Gluten sensitivity, GE Reflux, Obstructive apnea, Aspiration, Anemia, Thyroid disorders, Cervical spine, Seizures, Development-Behavior
Frequently Asked

Can Medical Conditions Impact Behavior Attention, Activity, Learning (Executive Function) in Children?
Sleep Disturbance in Children with DS

Frequently reported
Movement: restlessness, limb movement
Position: bent at waist, sitting up
Breathing: snoring, respiratory pause - not always reported

Problems with sleep initiation vs sleep maintenance
Worsened by GERD, Asthma, Nasal congestion

Must do overnight sleep study OR closely observe sleep between 1-6am
Sleep Disturbance Impacts Behavior and Executive Function

All mammals require sleep

Humans – spend up to 30% of their lives asleep

Cyclic occurrence of REM and NREM sleep stages

1st half- NREM and SWS (N3) predominate

2nd half- REM and N2 predominate

Critical for memory consolidation and daytime alertness
Sleep Cycles Throughout the Night

Stage 1 sleep and REM sleep (red) are graphed on the same level because their EEG patterns are very similar.
Sleep Initiation Disorders

Trouble falling asleep
Home environment - noise, activity, bedtime routines
Daytime napping - circadian shift
Computer/screen time - increased arousal
Behavioral - anxiety, verbal perseveration or “learned” behavior resistance/avoidance

Medications
Caffeine - chocolate, soft-drinks, energy drinks
Stimulants (ADHD), bronchodilators (asthma)
Antihistamines (URI), antidepressants (mood-anxiety)
Sleep Maintenance Disorders

**Trouble staying asleep**
- Room is too Hot or Dry
- Child is Thirsty or a Full Bladder

Restless, motorically active sleep
- Periodic limb movements - iron deficiency?
**Obstructive sleep apnea**

Medical – GE Reflux, Asthma, Nasal congestion
- Neurochemistry of DS/ID, ADHD, ASD, Anxiety
Treatment Options for Disrupted Sleep
NOT OSA

Behavioral
Bedtime routine – warm bath
Keep the room temperature cool

Medical
Treat respiratory infection, asthma, GE reflux
Avoid caffeine, stimulants, late meals
Supplement with iron if Ferritin levels are low
Melatonin or Benadryl
Obstructive Sleep Apnea in DS

Risk Factors
Small mandible (jaw bone) and crowded oral cavity
Narrow upper airway, narrow palate

Enlarged tonsils & adenoids (regrowth)
Nasal or sinus infection (congestion)
Decreased muscular tone – worse during REM sleep

Enlarged or widened tongue
Lingual tonsils

GE Reflux, Asthma
Treatment Options for Obstructive Sleep Apnea in DS

Treatments
Nasal decongestants: saline or steroid spray
Sleep position sideways or sitting up
Tonsillectomy
Positive Airway Pressure Mask

Management of GE Reflux, Asthma
Frequently Asked

Behavior Concerns in Children?
Behavior Concerns in Down Syndrome

70-80% do NOT have a significant behavior disorder
20-30% may have more significant behavior problems

Most children will display occasional problems with attention or behavior which concern parents or teachers

Many children exhibit traits that are characteristic of DS
- Overly social and affectionate
- Stubborn persistence
- Repetition and preferred routines
Neurobehavioral Disorder

Behaviors are atypical or maladaptive

Interfere with learning, socialization & daily function
May further jeopardize
  • personal safety
  • social acceptance
  • classroom placement
Externalizing & Internalizing Dimensions of Neurobehavioral Disorders

**Externalizing** - receives the most attention

Disruptive behavior interferes with group activities
Potentially harmful to self or others

**Internalizing** - somewhat tolerated

Social withdrawal & Repetitive behavior
Less disruptive to group activities
Difficult to engage in therapies & learning
Oppositional-Disruptive Behavior Spectrum

Spectrum of Severity: ADHD…ODD…DBD ~10% in DS

**Attention Deficit-Disorder** (Disregulation)
- Distractability, poor focus, low attention/vigilance
- Disorganized, low motivation

**Impulse Control Disorder** (Hyperactivity)
- Poor inhibitory control, overactive, good attention
- Learned behavior
- Safety concerns – RISK TAKERS
Oppositional-Disruptive Behavior Spectrum

Spectrum of Severity: ADHD...ODD...DBD ~10% in DS

**Disruptive Behavior Disorder** ( +/- ADHD)
- A pattern of behavior with episodic aggression or property destruction
- Learned behavior task escape/avoidance or social attention?
- Safety concerns

**Oppositional Defiant Disorder** ( +/- ADHD)
- A pattern of negativistic, defiant, non-compliant behavior
- Argumentative, annoys others, easily angered, episodic tantrums
- Low frustration tolerance
- Learned behavior
- Safety concerns
Disruptive Behavior Spectrum

Functional impairment in social, family, academic function
Present for > 4-6 months
Not exclusively part of a mood, anxiety or autistic disorder

Further Impacts
Travel
Medical-dental procedures
Physical movement and transitioning
Social-community events
Disruptive Behavior Spectrum
Additional Considerations

Medical concerns
Sleep disturbances
Illness, pain or discomfort
Hyperthyroid

Neurophysiologic attributes
Underlying anxiety or mood disorder (irritability)
Fatigue, tired, sleepy
Cognitive organization and style (rigid-inflexibility)
Internalizing features (withdrawal, perseveration, repetitive)
Sensory aversions (noise, chaos)
Disruptive Behavior Disorders

**Treatments**
Safety monitoring & supervision - running, climbing, jumping
Behavior management - recognize the setting, motivations, triggers, reinforcers
Sensory, avoid chaotic, loud environment
Teach relaxation techniques – massage, quiet hands
Functional communication - visual prompts, photographs, iPAD

Treat underlying medical conditions including sleep
Medications – to manage physiologic symptoms
Frequently Asked

Medical Conditions in Adults
How do they differ from children?
Figure 2   Mean, median, and 25th and 75th percentiles for age at death in persons with Down syndrome, 1900-2007. The mean and median age at death for persons with Down syndrome have increased significantly over the past 40 years.
Medical Guidance for Young Adults with DS

Based on physical findings and increased prevalence
*consensus needed*

**Medical Follow-up:** according to previous history

**Sustained Vigilance:**

**Younger Adult:** (<35yr) Sleep Apnea, Obesity, Mitral/Aortic Valve leak, Mood-anxiety disorder, Functional decline, Hypotension with dehydration (syncope)

**Ongoing Evaluations:** Weight management, Nutrition, Fitness-exercise, Thyroid, Vitamin D, Cholesterol, Vision, Hearing, Dental, Female: GYN care
Medical Guidance for Elderly Adults with DS

Based on physical findings and increased prevalence
*consensus needed*

**Medical Follow-up:** according to previous history

**Sustained Vigilance:**

**Elderly Adult: (>40yr)** Chewing-swallowing difficulties, Tooth loss, Weight loss, Aspiration pneumonia, Osteoporosis, Gait changes, Cervical spondylosis, Cataracts, Hearing loss, Seizures, Motor deterioration, Intellectual decline (AD-type)

**Ongoing Evaluations:** Nutrition and caloric supplements, Thyroid, Vision, Hearing, Dental, Self care skills, Safety and supervision in the home
Consensus guidelines for primary health care of adults with developmental disabilities

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Tom Cheetham, MD, CCFP  Brian Hennen, MD, CCFP  Elspeth A. Bradley, MBBS, PHD, FRCP, FRCPsych
Joseph M. Berg, MB, BCH, MSC, FRCPsych, FCCMG  Marika Korossy  Cynthia Forster-Gibson, MD, PHD
Maria Gitta, MA  Chrissoula Stavrakaki, MD, PHD, FRCP  Bruce McCreary, MD, FRCP  Irene Swift, MBBS

ABSTRACT

OBJECTIVE To develop practical Canadian guidelines for primary health care providers based on the best available evidence for addressing health issues in adults with developmental disabilities (DD).

QUALITY OF EVIDENCE Authors of background papers synthesized information from their own clinical experience, from consultations with other experts, and from relevant professional publications. Based on discussions of these papers at a colloquium of knowledgeable health care providers, a consensus statement was developed. Standard criteria were used to select guidelines for consideration and to rank evidence supporting them. Most evidence was level III.

MAIN MESSAGE People with DD have complex health issues, some differing from those of the general population. Adequate primary health care is necessary to identify these issues and to prevent morbidity and premature death. Physical, behavioural, and mental health difficulties should be addressed, and primary health care providers should be particularly attentive to the interactions of biological, psychological, and social factors contributing to health, since these interactions can easily be overlooked in adults with DD. Attention must also be paid to such ethical issues as informed consent and avoidance of harm. Developmental disabilities are not grounds for care providers to withhold or to withdraw medically indicated interventions, and decisions concerning such interventions should be based on patients' best interests.

CONCLUSION Implementing the guidelines proposed here would improve the health of adults with DD and minimize disparities in health and health care.
Colloquium on the Primary Health Care of Adults with Developmental Disability held Nov 7-11, 2005 Toronto

Evidence based
Expert opinion or consensus statement
Practical concise guidelines

Special mention of DS throughout
Inform clinical decision-making for PCPs within the existing Canadian Health Care system
Canadian Consensus Guidelines
Highlights

General Primary Care
• Review all medications every 90 days
• Medication monitoring
• Review of psychiatric indications for psychotropic medications

Physical Health
• Obesity, Exercise, Vision, Hearing
• Dental, Thyroid, GERD, Osteoporosis, Seizures

Behavioural & Mental Health
• Maladaptive behavior: aggression, self-injury, social withdrawal
• Psychiatric disorders: Anxiety, depression, compulsions, psychosis
• Acute behavioral crisis vs long-term treatment plan
• Risk for Dementia
Medical Management of Adults with Down Syndrome

Managing the care of adults with Down's syndrome

Obesity

Literature Review Summary

• The prevalence of obesity (BMI >30) in adults with DS is (~40%), higher than the prevalence in adults with other causes of ID (~25-30%).
• An estimated 150,000 x 0.4 = 60,000 DS adults living in the U.S. are obese.
• An additional 150,000 x 0.35 = 52,000 DS adults are overweight.
• Major health significance.
Obesity

Individuals at high risk:
• Female gender, adolescent onset, living in family home, having few recreational or leisure activities.
• The role of diet, exercise and other family factors has not been well explored.

Summary
• There is limited evidence about associated medical and behavioral comorbidities or the use of biomarkers in obesity.
• The number of DS subjects studied is (~1500); and study design is adequate for addressing some key questions (prevalence, screening, some risk-factors)
• Available evidence is of fair-good validity.
Risk Factors for Obesity

Female – puberty (estrogen) hormones
Metabolism – lower RMR
Hypothyroidism – not often the cause
Sleep apnea
Eating disorder, satiety signaling (leptin)
Medications - OBCP, Newer antipsychotics, SSRIs

Lifestyle
Low activity
Family BMI trend
Chronic Obesity

Associations
- Fatigue – lethargy, mood?
- Metabolic - hyperglycemia-hyperlipidemia? (rarely HTN, stroke)
- Sleep apnea - worsened
- Musculoskeletal - joint pain - accelerated degenerative changes?

Prevention-Treatment
- Controlled access to food - family
- Supervised exercise program - personal trainer
- Medically supervised weight loss (BMI >40 + multiple risk factors)

Medications: Is there a role?
- Thyroid management (levothyroxine or t4/t3)
- Eating as compulsive behavior (SSRI). Appetite suppression (stimulant)
- Hyperglycemia (hypoglycemics); Hyperlipidemia (statins)
Bones
Bone Density

Literature Review Summary

• Evidence for increased prevalence of osteoporosis in adults with Down syndrome compared to GP.
• Total number of DS subjects studied is ~400.
• 6 of 7 studies found an increased risk of osteoporosis compared to GP.
• 3 studies addressed osteoporosis in others with ID
• 2 of 3 studies found it was higher in DS, 1 of 3 found no difference.
• 1 of 6 studies found an increase in fractures compared to the GP, 5 of 6 did not.
Risk Factors for Low Bone Density

Bone matrix-composition early osteogenesis
Lower muscle tone/strength
Low physical activity, reduced sunlight exposure
Medications – anticonvulsants

Endocrine
- low freeT4 – hypothyroidism
- Amenorrhea
- low estrogen, testosterone – hypogonadism
- low Vitamin D – dietary intake/conversion, GI malabsorption
Preventing/Treating Osteoporosis

Adults > 35 yr  Review risk factors
  Monitor Vitamin D levels
  Estrogen and Testosterone - early menopause
  DEXA bone scan

Treatment
  Dietary intake and supplemental Vitamin D, Calcium
  Vit D3 (1000-2000IU) & Calcium (1000mg)
  Maintain physical activity & sunshine

Intervention
  Bisphosphonates: Fosamax, Actonel, Boniva?
  Hormone replacement? estrogen, testosterone
Mental Health
Adolescents & Adults Mental Health

Behavior or Mental Health concerns in 20-30%

- Change from previously established baseline
- Gradual onset or Intensification of pre-existing traits
- Post-traumatic onset - following physical illness or emotional stress
Major Depressive Episode & Down Syndrome

Frequency: Male = Female
Mood & Anxiety disorders: increases post-puberty
Grief, bereavement or adjustment reaction is NOT Major Depression

Is risk of MDE in DS Age associated?

- NOT a linear increase with ageing - peaks at 15-25yr
- NOT progressive

Prodrome of dementia?
(Amyloid peptide toxicity, glutamate/GABA/monoamine failure)
Major Depressive Episode & Down Syndrome

Warren et al. (1989) reported 5 DS subjects (17-38 years)

- Symptom-complex including cognitive decline, marked apathy, social withdrawal and psychomotor slowing
- Deterioration in adaptive skills from an established BL
- Distinguished subjects from those with AD-type dementia

Additional reports by

Myers and Pueschel (1995) 9 subjects (21-44 years)
Capone & Aidikoff (2011) 33 subjects (14-35 years)
Phenomenology of Functional Decline in Down Syndrome  Prasher, V  2002

Case reports (15-35 yr), peak occurrence ~22yr

- Severe deterioration in function following “typical” development for DS
- Loss in cognitive, speech, motor and adaptive skills – then plateau
- May not meet diagnostic criteria for depression or dementia
- Limited response to antidepressant medications
## Down Syndrome Case-Control Study

<table>
<thead>
<tr>
<th>Diagnostic Groups</th>
<th>DS Subjects</th>
<th>Functional decline</th>
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<tbody>
<tr>
<td>Major Depressive Episode (MDE)</td>
<td>N=19</td>
<td>13 (68%)</td>
</tr>
<tr>
<td>MDE + Psychosis</td>
<td>N=9</td>
<td>6 (66%)</td>
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<tr>
<td>Age at evaluation</td>
<td></td>
<td>21.0 ± 5 yr</td>
</tr>
<tr>
<td>Symptom Duration</td>
<td></td>
<td>12.8 ± 9.1 mo</td>
</tr>
<tr>
<td>Mentally Healthy DS (Controls)</td>
<td>N=9</td>
<td>0%</td>
</tr>
<tr>
<td>Age at evaluation</td>
<td></td>
<td>19.8 ± 7.1 yr</td>
</tr>
<tr>
<td>Thyroid Status</td>
<td>MDE</td>
<td>Controls</td>
</tr>
<tr>
<td>--------------------------------</td>
<td>--------------</td>
<td>--------------</td>
</tr>
<tr>
<td>Thyroid supplement</td>
<td>11/28 (39%)</td>
<td>4/9 (44%)</td>
</tr>
<tr>
<td>Euthyroid (TSH 0.5-5.5 ul U/ml)</td>
<td>27 (96%)</td>
<td>9 (100%)</td>
</tr>
<tr>
<td>Compensated (TSH &gt; 5.5 ul U/ml; normal T4)</td>
<td>1</td>
<td>0</td>
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<tr>
<td>Hypothyroid (low T4)</td>
<td>0</td>
<td>0</td>
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## Psychiatric Symptom Screener: Reiss Scales of Children’s Dual Diagnosis

<table>
<thead>
<tr>
<th>Subscale score</th>
<th>MDE</th>
<th>Controls</th>
<th>t-test</th>
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<tbody>
<tr>
<td></td>
<td>N=28</td>
<td>N=9</td>
<td></td>
</tr>
<tr>
<td>Anger</td>
<td>2.2 ± 1.7</td>
<td>1.7 ± 2.8</td>
<td>NS</td>
</tr>
<tr>
<td>Anxiety</td>
<td>2.4 ± 2.0</td>
<td>0.2 ± 0.4</td>
<td>&lt; .0001</td>
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<tr>
<td>Attention</td>
<td>2.2 ± 1.6</td>
<td>0.8 ± 1.0</td>
<td>&lt; .01</td>
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<tr>
<td>Autism</td>
<td>2.5 ± 1.8</td>
<td>0.1 ± 0.3</td>
<td>&lt; .0001</td>
</tr>
<tr>
<td>Conduct</td>
<td>0.6 ± 0.8</td>
<td>0.4 ± 1.0</td>
<td>NS</td>
</tr>
<tr>
<td>Depression</td>
<td>4.6 ± 2.0</td>
<td>0.7 ± 1.4</td>
<td>&lt; .0001</td>
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<tr>
<td>Poor Self-Esteem</td>
<td>1.4 ± 1.9</td>
<td>1.0 ± 1.3</td>
<td>NS</td>
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<tr>
<td>Psychosis</td>
<td>4.3 ± 2.2</td>
<td>0.3 ± 0.7</td>
<td>&lt; .0001</td>
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<tr>
<td>Somatoform</td>
<td>1.2 ± 1.5</td>
<td>1.2 ± 2.1</td>
<td>NS</td>
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<tr>
<td>Withdrawn</td>
<td>5.8 ± 2.4</td>
<td>0.9 ± 1.4</td>
<td>&lt; .0001</td>
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<tr>
<td>Other Behavior</td>
<td>2.9 ± 1.9</td>
<td>1.0 ± 1.9</td>
<td>.01</td>
</tr>
<tr>
<td>Reiss Total</td>
<td>30.4 ± 10.9</td>
<td>8.4 ± 8.5</td>
<td>&lt; .0001</td>
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## Maladaptive Behavior: Aberrant Behavior Behavior Checklist

<table>
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<tr>
<th>Subscale</th>
<th>MDE</th>
<th>Controls</th>
<th>t-test</th>
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<tbody>
<tr>
<td>N=28</td>
<td>N=9</td>
<td>&lt;.01</td>
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<tr>
<td>Irritability</td>
<td>8.8 ± 7.2</td>
<td>2.6 ± 3.7</td>
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<tr>
<td>Hyperactivity</td>
<td>9.8 ± 8.7</td>
<td>4.9 ± 3.9</td>
<td>&lt;.05</td>
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<tr>
<td>Lethargy</td>
<td>21.0 ± 10.0</td>
<td>3.1 ± 4.1</td>
<td>&lt;.0001</td>
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<tr>
<td>Stereotypy</td>
<td>6.9 ± 5.1</td>
<td>0.7 ± 1.1</td>
<td>&lt;.0001</td>
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<tr>
<td>Speech</td>
<td>3.1 ± 3.0</td>
<td>2.0 ± 3.3</td>
<td>NS</td>
</tr>
<tr>
<td>ABC Total</td>
<td>49.6 ± 24.1</td>
<td>13.3 ± 12.0</td>
<td>&lt;.0001</td>
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ABC Maladaptive Behavior

“Spider-Graph” Behavior Composite MDE vs Controls
Does Sleep Apnea Contribute to Symptoms of Depression?
## Sleep Screener: Sleep Disturbance Scale

<table>
<thead>
<tr>
<th>Subscale</th>
<th>MDE</th>
<th>Controls</th>
<th>t-test</th>
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<tr>
<td><strong>N=19</strong></td>
<td><strong>N=6</strong></td>
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<td></td>
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<tr>
<td>Initiation</td>
<td>19.4 ± 6.7</td>
<td>11.5 ± 2.4</td>
<td>&lt; .01</td>
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<tr>
<td>Maintenance</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Breathing</td>
<td>6.4 ± 3.1</td>
<td>3.3 ± 0.5</td>
<td>&lt; .05</td>
</tr>
<tr>
<td>Arousal</td>
<td>3.3 ± 0.7</td>
<td>3.0 ± 0</td>
<td>NS</td>
</tr>
<tr>
<td>Transition</td>
<td>10.0 ± 3.4</td>
<td>8.0 ± 2.4</td>
<td>NS</td>
</tr>
<tr>
<td>Somnolence</td>
<td>12.1 ± 5.0</td>
<td>9.0 ± 2.4</td>
<td>NS</td>
</tr>
<tr>
<td>Hyperhydrosis</td>
<td>2.7 ± 1.5</td>
<td>2.0 ± 0</td>
<td>NS</td>
</tr>
<tr>
<td>SDS Total Score</td>
<td>53.9 ± 11.8</td>
<td>36.8 ± 3.6</td>
<td>&lt; .01</td>
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</tbody>
</table>
# Respiratory Events & Oxygenation Data

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<thead>
<tr>
<th></th>
<th>MDE</th>
<th>Controls</th>
<th>t-test</th>
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</thead>
<tbody>
<tr>
<td><strong>Respiratory Events</strong></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Apnea/Hypopnea Index (n/hr)</td>
<td>18.8 ± 14.7</td>
<td>4.5 ± 3.8</td>
<td>.001</td>
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<tr>
<td>REM-Apnea/Hypopnea Index (n/hr)</td>
<td>35.8 ± 35.9</td>
<td>11.9 ± 10.2</td>
<td>.01</td>
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<tr>
<td>Respiratory Disturbance Index (n/hr)</td>
<td>24.3 ± 24.6</td>
<td>7.3 ± 7.3</td>
<td>&lt; .01</td>
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<tr>
<td>Central Apneas (n/TST)</td>
<td>4.9 ± 16.0</td>
<td>2.3 ± 4.0</td>
<td>NS</td>
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<tr>
<td><strong>Oxygenation</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean Sa02 during sleep (%)</td>
<td>95.1 ± 1.4</td>
<td>96.3 ± 2.7</td>
<td>NS</td>
</tr>
<tr>
<td>Sa02 nadir during sleep (%)</td>
<td>83.7 ± 8.4</td>
<td>92.0 ± 6.7</td>
<td>&lt; .01</td>
</tr>
<tr>
<td>Sa02 &lt; 90% (TST%) [minus outlier ]</td>
<td>3.7 ± 5.2</td>
<td>3.9 ± 11.6 [0.0]</td>
<td>NS [001]</td>
</tr>
</tbody>
</table>
# Sleep Apnea: Severity & Tonsillectomy Status

<table>
<thead>
<tr>
<th>Mental Health Diagnosis</th>
<th>Mod-Severe apnea</th>
<th>Mild apnea</th>
<th>No apnea</th>
<th>Previous tonsil removal</th>
<th>% with Sleep Apnea</th>
</tr>
</thead>
<tbody>
<tr>
<td>Major depressive episode (MDE) N=28</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>86%</td>
</tr>
<tr>
<td>No mental health diagnosis N=9</td>
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<td></td>
<td></td>
<td></td>
<td>44%</td>
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<tr>
<td>Previous tonsillectomy</td>
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<td></td>
<td></td>
<td></td>
<td>74%</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th></th>
<th>N=16</th>
<th>N=12</th>
<th>N=9</th>
<th>N=15</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Major depressive episode (MDE) N=28</td>
<td>15 (54%)</td>
<td>9 (32%)</td>
<td>4 (14%)</td>
<td>12 (43%)</td>
<td>86%</td>
</tr>
<tr>
<td>No mental health diagnosis N=9</td>
<td>1 (11%)</td>
<td>3 (33%)</td>
<td>5 (55%)</td>
<td>3 (33%)</td>
<td>44%</td>
</tr>
<tr>
<td>Previous tonsillectomy</td>
<td>6 (38%)</td>
<td>5 (41%)</td>
<td>4 (45%)</td>
<td>15 (40%)</td>
<td>74%</td>
</tr>
</tbody>
</table>
Managing Co-occurring Sleep & Mental Health Disorders

Goals - What are we treating? in what priority?

• Respiratory disturbance with hypoxemia
• Sleep fragmentation with arousals

• Depression or frontal lobe dysfunction?
• Dementia or pseudo-dementia syndrome?

• A disease model needs to be worked out
Managing Sleep & Mental Health Disorders

Step-wise referral for separate medical, psychiatric, pulmonary/sleep & ENT evaluation rarely proceeds efficiently

• Lengthy delays before proper treatment can commence
• Requires a concerted and creative approach to diagnostic evaluation and management
• Consider sleep-team for complex disorders?

• Can we reliably get a full ON PSG?
• What about home video monitoring /02 saturations
Managing Sleep & Mental Health Disorders

Prioritizing Management - How hard do we try?
What is the family’s willingness to try?

- Sleep positioning techniques
- CPAP/BiPAP adherence
- Behavioral desensitization or anti-anxiety/sleep medications?
- Surgery T&A, lingual tonsils or UPPP
- Weight reduction is not usually easy or effective
- Psychotropic medications maybe recommended for mood or psychotic symptoms but can contribute to weight gain
Current Estimate 6-8 Million Persons w/ DS Worldwide
Health Care for All Persons with Down Syndrome

Improvements in health care have been “uneven”

- Limited by access to medical knowledge, resources or implementation of best practices
- Challenge of providing care to complex individuals
Health Care Challenges Ahead

Family & NGOs & Advocacy Organizations

• Educate families to recognize/anticipate medical needs
• Disseminate information to community Primary Care Providers
• Stay up-to-date - books, websites, conferences
• Create NEW services
Health Care Challenges Ahead

Health Care Providers

• Creation of Adult guidelines requires coordination & consensus
• Implementation of guidelines requires time & resources
• Access to specialty consultation is required
• Complex conditions including behavior and mental health
• Dementia and End-of-Life decision making & care
The END Game

Things will be all right in the END...

..If it is NOT yet right, then it’s not the END

Sonny Kapoor
Thank You