Awareness, Diagnosis, and Management of Osteoporosis in Adults with Developmental Disabilities

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Health, Bones and Vitamin D

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IFCD.org - Caring for People with Disabilities
Why Osteoporosis is a Major Problem in DD Patients

• Men and women with developmental disabilities receive sub-standard health care (and perhaps, neglect)

• In particular, preventative services are under used in this population

• People with disabilities continue to face barriers to receiving health care ($1^0 \& 2^0$)
  - Access; Attitudes of providers
  - Lack of information about health care needs
The Number of Americans with Chronic Conditions is Expected to Rise

33 Million in 2010

28 Million in 1995

39 Million in 2020

In 1995, 99 million people had chronic conditions

28 million (28%) had limitation in major activity

It is projected that in 2020, 134 million people will have chronic conditions

39 million (29%) will have limitation in major activity
But Pool of Potential Caregivers are Shrinking
Economics of Care-Giving

• The value of services family caregivers provide for “free” is estimated to be over $290 billion/yr – twice as much as is actually spent on homecare and nursing home services.

• Of estimated 2.7 million Americans who need assistive technology such as wheelchairs, 64% can’t afford it.

• In patients with DD, this is almost 100%
Incidence of Developmental Disabilities in US

- 26 million people with disabilities in US
- Many types of disabilities (ranging from mental health issues to multiple sclerosis)
- If a third has OP, this is an additional 8 million people with OP in USA alone (not accounted for)
- Over 500,000 people in US, over the age of 60 has developmental disabilities
- This number projected to be doubled by 2030
Time Line and Phases of Illnesses

Patients with Chronic Disabilities, it is a Continuum, with a shorter life expectancy

Time Line and Phases of Illness

Prediagnosis with symptoms

Initial adjustment period

Chronic "long haul"

Preterminal

Mourning and resolution of loss

Why Osteoporosis is a Major Problem in DD Patients

• The incidence of osteoporosis and fracture rates are over ten-times higher in developmental disabilities patients

• Diagnostic and the preventative services are grossly under used in this population

• They have multiple barriers to receiving health care ($1^0 \& 2^0$)
  – Access; Attitudes of providers
  – Lack of information about health care needs
Osteoporosis & Disabilities: What is the Problem?
Problems:

- Pain & difficulty
- Loss of independence
- Depression
- Loss of height
- Compression of chest & abdomen
- Treatment with inappropriate medications
- Increase morbidity and mortality
Identification of High-Risk Patients: Role of Age and Gender

Osteoporotic (Fragility) Fractures

Aging

High bone turnover

Menopause

Bone loss

Propensity to fall

Fractures

Low peak bone mass

Hormonal environment

Developmental abnormalities

Genetic factors

Exercise

Nutrition
Fracture Risk Doubles With Every S.D. Decrease in BMD

Relative Risk for Fracture

Bone Density (T-score)
History of Bone Density Testing
New Jersey, 2007
What will be Discussed

• What is osteoporosis?
• Why should we worry about it?
• What are the issues facing with development disabilities?
• How can you make a diagnosis?
• What has been done for patients with disabilities?
• What can we do to improve the situation?
What is Osteoporosis?
Osteoporosis - Definition

“Skeletal disorder characterized by compromised bone strength, predisposing to increased risk of fractures” *

* National Institutes of Health Consensus Development Panel on Osteoporosis Prevention Diagnosis, and Therapy
According to the National Osteoporosis Foundation (NOF):

- 44 million people in the U.S. have low BMD (< 1g/cm\(^2\))
  - 11 million of those have osteoporosis
  - 35 million have osteopenia and are at risk of for osteoporosis

- Projection: By 2020 there will be 15 million Americans with osteoporosis.
Osteoporosis is a Largely Under-diagnosed Condition

- Often diagnosed after a fracture
- Patient are usually asymptomatic prior to a fracture
- Low bone mineral density (BMD) is considered the "disease"
- Fragility fractures are the "consequences" of the disease
Osteoporosis affects the quality of bone.
All Patients Receiving any of the Following Should be Advised of the Potential Risks of Osteoporosis

- Any chronic disability
- Gastrectomy / stapling
- GI pathology or surgery
- Any fragility fracture
- Glucocorticoid therapy
- Anti-convulsants
- Transplants / immuno-suppressant
- Depo-Provera, Lupron, etc.
Special Problems in Patients with Developmental Disabilities

- Intellectual (e.g., Down’s syndrome)
- Other mental disabilities
- Physical & cognitive disability
- Immobility
- Excessive medication use
  - ~40% taking anti-epileptics
  - ~50% taking anti-psychotic medication
- Low vitamin D levels (high incidence)
Special Problems in Patients with Developmental Disabilities

• High incidence of non-traumatic fractures (8-18%)
• Low bone density (40-80%)
  — Osteopenia (50%) & Osteoporosis (30%)
• Immobility & seizure disorders
• Medication use (anti-epileptics)
• Low vitamin D level (↓ sun exposure)
## Osteoporosis in Normal Persons vs. Patients with Developmental Disabilities

<table>
<thead>
<tr>
<th>Condition</th>
<th>Normal Population</th>
<th>Developmental disabled patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Undiagnosed</td>
<td>40%</td>
<td>90%</td>
</tr>
<tr>
<td>Prevalence of:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Low bone density</td>
<td>15%</td>
<td>70%</td>
</tr>
<tr>
<td>Vitamin D deficiency</td>
<td>40%</td>
<td>90%</td>
</tr>
<tr>
<td>Falls</td>
<td>1</td>
<td>20-times higher</td>
</tr>
<tr>
<td>Fracture rates</td>
<td>1</td>
<td>10-times higher</td>
</tr>
<tr>
<td>Life expectancy</td>
<td>70-80 yrs</td>
<td>20-50 yrs</td>
</tr>
</tbody>
</table>
Some Facts About Osteoporosis:

• In older women, incidence of osteoporotic fractures is greater than incidence of myocardial infarction, stroke, and breast cancer combined.

• Can appear in both men and women; adults & children

• Osteoporosis and fractures are not solely the outcome of post-menopausal status in women
Importance of Risk Factors in the Diagnosis of Osteoporosis

Risk factors are additive in predicting fractures
Combining BMD With Clinical Risk Factors

U.S. Fracture Incidence:

- Approx. 1.5 million people per year have osteoporosis-related fragility fractures:
  - 700,000 spine
  - 300,000 hip
  - 250,000 wrist
  - 300,000 other

- Economic costs, including rehab, hospitalization, and nursing home care:
  - Direct cost in 1995 - $13.8 billion
  - 2003 -$17 billion
  - 2008 -$21 billion

www.NOF.org
Health Costs of Osteoporosis-Related Falls and Fractures

- 1.5 million osteoporosis-related fractures in the U.S. annually
- In New Jersey osteoporosis-caused bone fractures (~36,630 each year), cost $496 million annually
- Hospitalization due to Hip fractures cost between $18,000-$26,912. Double by 2020 to an estimated $60,000.
- Falls the second leading cause of injury-related New Jerseyans 65 years and older
- >90 percent of hip fractures are associated with osteoporosis and nine out of 10 are the result of a fall.
- Between 2000 and 2005, fatality from falls among New Jerseyans 65 to 84 years old and 85 years and older has nearly doubled.
“Low Bone Mass” of Osteoporosis Have Multiple Causes

- Vitamin D deficiency
- Endocrine disorders:
  - Glucocorticoid therapy, Cushing’s disease
  - Hyperparathyroidism
  - Hyperthyroidism
- Drug toxicity (anti-epileptic drugs)
- Immobility
- Hypogonadism (in men and women)
Populations at Risk of Developing Osteoporosis & Fractures

- Glucocorticoid (steroid) use
- Eating disorders
- Persistent amenorrhea
- Low sex-hormone levels
- Thyroid/parathyroid diseases
- Frequent fallers
- Developmental disabilities
70-Year-old Woman with Newly Diagnosed Temporal Arteritis, Treated with Prednisone

Baseline

One Year Later

A preventable cause of OP
Common sites of fractures in developmentally disabled individuals:

- Not different from normal population
- Cause is often undeterminable
- May lead to additional disabilities and premature death
Hip Fracture Complications

- 30% excess mortality within the year from the fracture (nearly 65,000 women die)
- 50% survivors are incapacitated
- 22% require long-term nursing home care
- Failure to diagnose and treat osteoporosis
- In 50% of the survivors, the second hip fracture occurs within four years

Diagnosis of Osteoporosis

• Begins with a DXA scan, which is normally straightforward in the general population

• Adults with DD may present with contractures, deformities, and movement disorders that may confound analysis of BMD.

• Regions of Interest (ROI’s) may need to be adjusted for valid interpretation.
Bone density is the single best predictor of bone strength and fracture risk.

DXA is a major breakthrough in diagnosis and treatment of osteoporosis:
- Diagnose osteoporosis prior to fractures
- Assess an individual’s risk for fracture
- Monitor therapy
Example of a DXA Bone Density Testing Machine
Bone density diagnosis is based on result at lumbar spine, total hip, and femoral neck sites only. Other sites (Ward’s triangle, etc.) are not valid predictors of fracture risk.
Treatment of Osteoporosis

• May require additional consideration of multiple co-morbid conditions

• No studies have been done to demonstrate efficacy of “bone-building” pharmaceuticals in persons with DD with low bone density

• Few medical specialists in treating metabolic bone disorders that are experienced with the health problems in DD population
Treatment of Osteoporosis

- There are no established standards of care for the evaluation and management of osteoporosis or fractures in DD population.

- More clinical data, research, and guidance are needed.

- IFCD Foundation is promoting such.
BMD Data

Analysis of BMD from individuals who had baseline and follow-up DXA scans suggests:

- Calcium and Vitamin D alone are not sufficient to improve BMD
- Low BMD is more prevalent in the hips than in the spine
- Bisphosphonates appear to be more effective for low BMD of the spine, but very little effect on the hips
- Vitamin D treatment is highly cost-effective
Treatment of Osteoporosis

• Even though it may increase the BMD, it is possible that none of the available FDA-approved anti-osteoporosis therapies will prevent fractures significantly in this community.

• In addition to low BMD, fractures in DD patients are due to a different set of reason than the normal population:
  – Excessive use of drugs and drug interactions
  – Poor balance, coordination and ↑ falls
  – Aggression, etc.
Guidelines for BMD Measurements

• Most disabled patients aught to have BMD testing

• Should have baseline BMD prior to, or early as possible of starting therapy

• Follow-up annually until bone mass stabilizes, and then every 2-3 years

• Measuring one site alone may miss rapid loss of BMD in other site (spine vs. hip)
Very Little Research has been done in Patients with Development Disabilities
Osteoporosis Research in Patients with Developmental Disabilities

- Tyler et. al., Mental retardation, 38: 316-321, 2001
- Schrager, S., J Women’s Health, 13: 431-437, 2004
- Schrager S., Mental retardation, 44: 203-211, 2004
Special Problems: Other Considerations

- As people age, their BMD decrease, while their risk of falls increase.
- Many people with disabilities take medications that may predispose to falls, e.g., agent with anti-cholinergic side-effects, as prescribed for incontinence, drooling, neuropathy, allergies, or psychiatric drugs.
Special Problems: Anti-convulsant Medications

- Impair vitamin D metabolism
  - Phenytoin, Cabamezapine, Valproic acid
- Toxicity to osteoblasts:
  - Phenytoin, Cabamezapine,
- Newer anti-epileptic agents (↓Vitamin D)
  - Topiramide, Gabapentin, Lamotrigine, Ethosuximide
- Poor nutrition
Special Problems: e.g., Down’s syndrome

- Lower peak bone mass
- Low bone density (up to 87%)
- Lower muscle tone
- Low vitamin D levels
- Increase falls and injuries
- Anti-convulsion medications
- Lives longer now
- High incidence of fractures
Special Problems: Spinal Cord Injuries & Stroke

• Immobility and lack of gravity may induce rapid bone loss in lower extremities (50% loss in 4 years)
• High incidence of fractures
• Lower muscle tone & low serum vitamin D levels
• Increase falls and injuries
Special Problems: Multiple Sclerosis and MG

- 350,000 people are affected with MS
- 75% of them are women
- Lower BMD, and
- High prevalence of vitamin D deficiency
- Poor muscle tone
- Increase falls and injuries
- High fractures rates
Special Considerations: Therapeutic Options

- Inability to take appropriate oral therapy e.g., oral bisphosphonates
- Annual or 3-monthly intravenous treatment with bisphosphonates
- Drug interactions (e.g., anti-convulsant)
- Immobility – difficulties with exercise
- Loss of mechanical stresses
- Frequent use of glucocorticoids (e.g., in MS, myasthenia gravis)
Fall and Injury Prevention

- Institutionalized and bed-bound patients are at much higher risk of osteoporosis and associated fractures
- Many of these patients eventually develop pathological fractures with minor or no trauma
- Unfortunately, many law suites have been filed inappropriately (i.e., handling of these patients vs. OP as a cause of the fracture)
Fall and Injury Prevention (Very Important)

- Increase muscle strength
- Environmental modification
- Hip protective pads
- Attention to vision & hearing
- Avoid medications that affect balance and co-ordination
We do not know whether bone-active pharmaceutical agents are effective in the DD individuals.

Early diagnosis, correcting vitamin D deficiency, and fall prevention are the most cost-effective ways to address this disease in patients with developmentally disabilities.
Prevention of Fractures
Each Patient is Unique

- **Diagnosis-DXA:** Documentation of osteoporosis or low bone mass
- **Blood tests:** Biochemical tests to exclude secondary causes of bone loss
- **Special care:** Education staff on how to handle these patients
- **Therapy:** Initiation of appropriate therapy such as vitamin D
Principals of Management of Osteoporosis – What Can We Do?

- Identification and treatment of secondary causes of osteoporosis
- Ensure adequate calcium and vitamin D, and weight-bearing exercise
- Commencing and maintenance of an cost-effective therapy
- Follow-up and monitoring of patients
Prevention/Treatment of Osteoporosis: General Recommendations

• Calcium: ~1,500 mg/day from the “diet and supplements”
• Vitamin D ~ 2,000 to 4,000 IU/day (or 50,000 IU, twice a month)
• Weight-bearing exercise
• Avoidance of tobacco, & ↓ alcohol intake
• Minimize medications that adversely affecting skeleton
• Fall prevention program
Treatment Goal for Osteoporosis

Fracture Reduction

Improving bone density and maintaining the structural integrity is important

and

Falls and injury prevention is critical in preventing fractures
Agents used in the Prevention and Treatment of Osteoporosis (FDA Approved therapies)

Adequate Calcium (~1,500 mg/day) and Vitamin D (~1,000 IU/day) intake

• Estrogen (± Progesterone)
• Bisphosphonates (Alendronate, Risedronate, Ibandronate, zoledronic acid)
• Calcitonin (a week agent)
• Selective estrogen receptor modulating agents (SERMs) (Raloxifene)
• PTH (1-34) - Teriparatide
Osteoporosis:
- Bone loss and fractures are preventable, and
- Irrespective of the degree of bone loss, condition or the age,
- Osteoporosis is treatable
In the future, health care professionals taking care of disabled patients will also need to be magicians, so that they will be able to check multiple things including bones without x-rays!
Summary

- Prevention is better than cure
- Identifying and eliminating secondary causes of bone losses
- Drug treatment may not be the answer
- Think – Nutrition, Calcium, Vitamin D
- Weight-bearing exercises
- Eliminate risk factors
- Prevent falls & injuries
Summary

1. Prevention is better than cure
2. Adolescence - window of opportunity
3. Nutrition
4. Calcium/Vitamin D
5. Exercise
6. Eliminate risk factors
7. HRT/Bisphosphonates/SERMS
8. Prevent Falls
Discussion

Other developmental centers should consider having a “Healthy Bones Initiative” program at their facilities.

With adequate identification, research, intervention, and data collection at the state-wide level, perhaps this could become a health topic in the national arena.
Don’t forget to talk to your Patient and their family members about his/her Risks of Osteoporotic Fractures

Fracture:
- Is osteoporosis
- Can be prevented
- Leads to more fractures
Vertebral Fractures: Accumulation of Cascade Fracture

Fractures are preventable
Whoever we are, wherever we live, we are human beings