

Introduced by Senator Fuller

June 3, 2014

Senate Concurrent Resolution No. 126—Relative to X-linked adrenoleukodystrophy.

LEGISLATIVE COUNSEL'S DIGEST

SCR 126, as introduced, Fuller. Adrenoleukodystrophy Awareness Month.

This measure would memorialize the month of November 2014 as Adrenoleukodystrophy Awareness Month.

Fiscal committee: no.

1 WHEREAS, X-linked adrenoleukodystrophy (X-ALD) is one
2 of a group of genetic disorders called the leukodystrophies that
3 cause damage to the myelin sheath, an insulating membrane that
4 surrounds nerve cells in the brain; and

5 WHEREAS, Women have two X chromosomes and are the
6 carriers of the disease, but men are most severely affected because
7 men only have one X chromosome and lack the protection provided
8 by an extra X chromosome; and

9 WHEREAS, The loss of myelin and the progressive dysfunction
10 of the adrenal gland are the primary characteristics of X-ALD; and

11 WHEREAS, X-ALD affects an estimated one in every 20,000
12 boys, which is roughly 13,600 people in the United States,
13 including boys like Jeremy Hill Jr. of Bakersfield; and

14 WHEREAS, While nearly all patients with X-ALD suffer from
15 adrenal insufficiency, also known as Addison's disease, the
16 neurological symptoms can begin either in childhood or adulthood;
17 and

1 WHEREAS, The childhood cerebral form is the most severe,
2 with onset between ages four and 10; and

3 WHEREAS, The most common symptoms are behavioral
4 changes, including abnormal withdrawal or aggression, poor
5 memory, and poor school performance; and

6 WHEREAS, Other symptoms include visual loss, learning
7 disabilities, seizures, poorly articulated speech, difficulty
8 swallowing, deafness, disturbances of gait and coordination,
9 fatigue, intermittent vomiting, increased skin pigmentation, and
10 progressive dementia; and

11 WHEREAS, The milder adult-onset form is known as
12 adrenomyeloneuropathy (AMN), which typically begins between
13 21 and 35 years of age, and has symptoms that include progressive
14 stiffness, weakness or paralysis of the lower limbs, and ataxia; and

15 WHEREAS, Almost one-half the women who are carriers of
16 X-ALD will develop a milder form of AMN, but almost never will
17 develop symptoms seen in boys with X-ALD; and

18 WHEREAS, Treatment with adrenal hormones can be lifesaving,
19 and symptomatic and supportive treatments for X-ALD include
20 physical therapy, psychological support, and special education;
21 and

22 WHEREAS, Newborn screening is effective in catching and
23 preventing the negative effects of X-ALD and is relatively
24 inexpensive; and

25 WHEREAS, The prognosis for patients with childhood cerebral
26 X-ALD is generally poor due to progressive neurological
27 deterioration, unless bone marrow transplantation is performed
28 early; and

29 WHEREAS, Recent evidence suggests that a mixture of oleic
30 acid and erucic acid, known as “Lorenzo’s Oil,” administered to
31 boys with X-ALD prior to the onset of symptoms can prevent or
32 delay, but not stop, the appearance of the childhood cerebral form
33 of X-ALD; and

34 WHEREAS, Death usually occurs between one and 10 years
35 after the onset of symptoms of childhood cerebral X-ALD, and in
36 adult-onset AMN deterioration will progress over decades; and

37 WHEREAS, November, the month of Jeremy Hill Jr.’s birthday,
38 was chosen to acknowledge the struggles of Jeremy Hill Jr., his
39 parents, Jeremy and Debra Hill, and his sister, Meagan Hill, in
40 fighting X-ALD; now, therefore, be it

1 *Resolved by the Senate of the State of California, the Assembly*
2 *thereof concurring,* That the Legislature does hereby proclaim the
3 month of November 2014 as Adrenoleukodystrophy Awareness
4 Month; and be it further

5 *Resolved,* That the Secretary of the Senate transmit copies of
6 this resolution to the author for appropriate distribution.