

# Mortality among California's Sickle Cell Data Collection Cohort, 2005-2014

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## BACKGROUND AND OBJECTIVES

- Although life expectancy has improved dramatically in the last 40 years for those living with sickle cell disease (SCD), this disorder continues to have high mortality at lower ages than the general population.
- Information about age of death, cause of death, and related factors among those living with SCD is typically limited to data collected from a clinical setting or from single source administrative data (such as death certificates).
- These methods omit individuals who did not receive care in settings that collect and publish data on their patients with SCD, or patients without SCD listed as a cause of death, respectively.
- CDC has implemented the Sickle Cell Data Collection (SCDC) program in California and Georgia to establish true population-based surveillance systems at the state level in an effort to address such gaps in knowledge.
- SCDC data allow investigation of health outcomes for patients with SCD who receive care in all settings.

## METHODS

- Hospital admissions, emergency department (ED), and Medicaid claims data for the years 2004 through 2015 were obtained by the SCDC programs.
- Patients with a SCD International Classification of Diseases (ICD) 9 or 10 code having three or more healthcare encounters within any five-year period are included in the cohort presented here.
- This cohort was linked by social security number and date of birth to the state's death records for the same period to identify deaths.
- The most common causes of death (COD) were determined using the 'underlying COD' variable and categorized based on the ICD 10 coding scheme.
- A SCD COD in the death record was determined by a search of underlying and contributing (where available) COD (ICD 10 codes).

## RESULTS

- There were 5,248 persons meeting the case definition of SCD identified in the hospital, ED, and claims data during the 12-year time frame.
- Of these, matching death records were found for 543 of these patients (10%).
- The mean age of death for the SCD patients was 44.2 years (median 45.7 years; SD 16.0 years).
- 39% of deaths occurred among those younger than 40 years of age.
- 75% of deaths happened in a hospital setting.
- Table 1 shows age, sex, setting and COD values.

## RESULTS

Figure 1: Age and Sex of Deceased Californians with SCD 2004-2015, n = 543

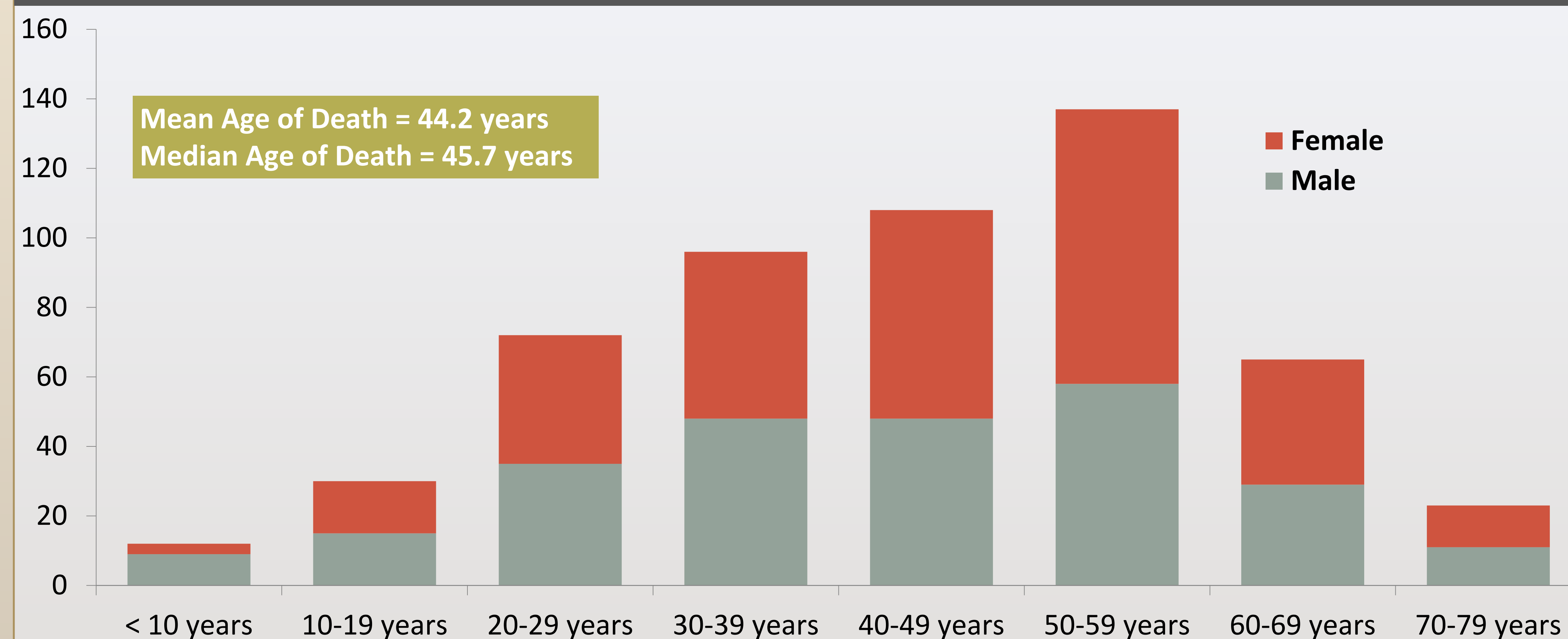


Figure 2: Setting of Deaths of Californians with SCD 2004-2015, n = 543

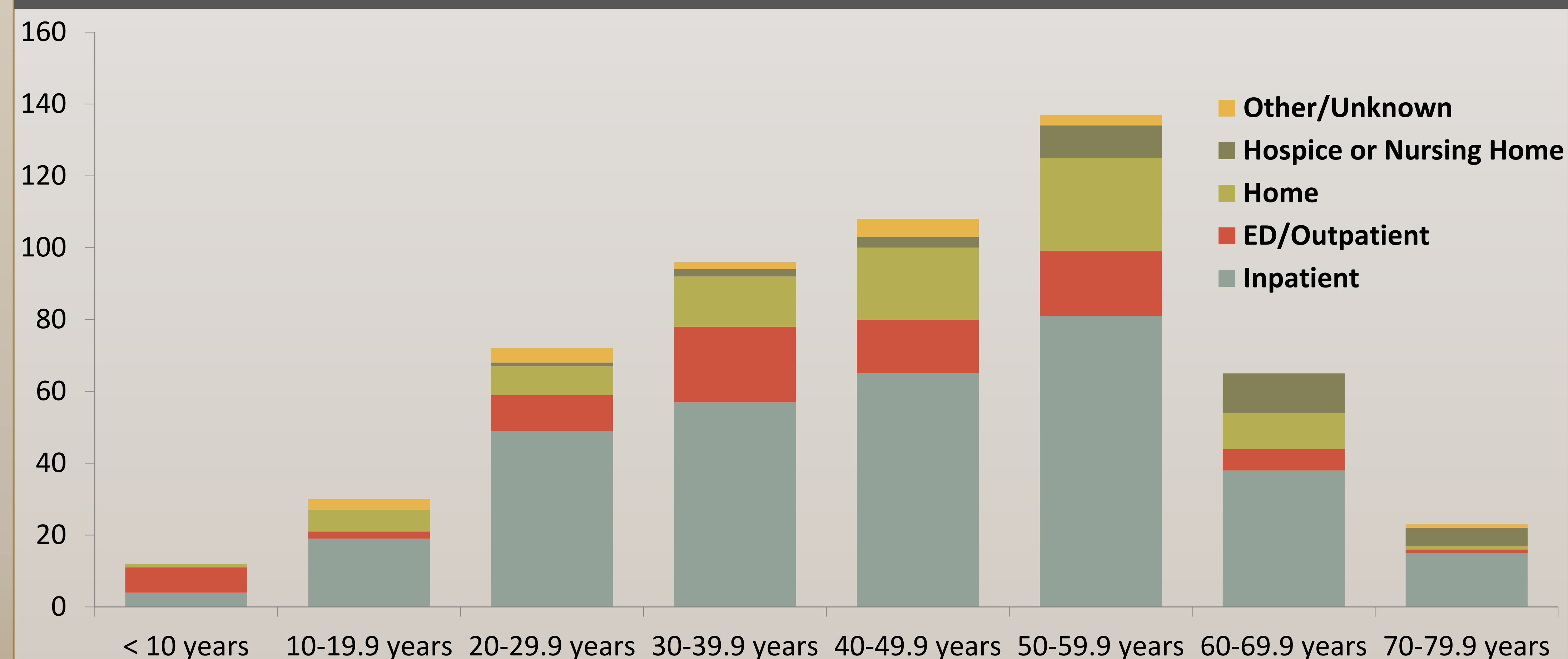


Table 1: Mortality among Californians with SCD 2004-2015 (%)

	< 10 years	10-19 years	20-29 years	30-39 years	40-49 years	50-59 years	60-69 years	70-79 years	Total (col. %)
<b>Sex</b>									
Male	9 (75)	15 (50)	35 (49)	48 (50)	48 (44)	58 (42)	29 (45)	11 (48)	253 (47)
Female	3 (25)	15 (50)	37 (51)	48 (50)	60 (56)	79 (58)	36 (55)	12 (52)	290 (53)
<b>Site of Death</b>									
Inpatient	4 (33)	19 (63)	49 (68)	57 (59)	65 (60)	81 (59)	38 (58)	15 (65)	328 (60)
ED/Outpatient	7 (58)	2 (7)	10 (14)	21 (22)	15 (14)	18 (13)	6 (9)	1 (4)	80 (15)
Home	1 (8)	6 (20)	8 (11)	14 (15)	20 (19)	26 (19)	10 (15)	1 (4)	86 (16)
Hospice/Nursing	0 (0)	0 (0)	1 (1)	2 (2)	3 (3)	9 (7)	11 (17)	5 (22)	31 (6)
Other/Unknown	0 (0)	3 (10)	4 (6)	2 (2)	5 (5)	3 (2)	0 (0)	1 (4)	18 (3)
<b>Cause of Death</b>									
Hematologic	5 (42)	5 (17)	37 (51)	37 (39)	50 (46)	53 (39)	16 (25)	3 (13)	206 (38)
Circulatory	0 (0)	1 (3)	11 (15)	18 (19)	12 (11)	35 (26)	21 (32)	9 (39)	107 (20)
External	2 (16)	6 (20)	9 (13)	15 (16)	7 (6)	10 (7)	2 (3)	0 (0)	51 (9)
Neoplasm	0 (0)	8 (27)	1 (1)	3 (3)	3 (3)	7 (5)	8 (12)	1 (4)	31 (6)
Infectious	3 (25)	2 (7)	3 (4)	3 (3)	9 (8)	6 (4)	4 (6)	0 (0)	30 (6)
Digestive	1 (8)	2 (7)	2 (3)	4 (4)	6 (6)	13 (9)	2 (3)	0 (0)	30 (6)
Other	1 (8)	6 (20)	9 (13)	16 (17)	21 (19)	13 (9)	12 (18)	10 (43)	88 (16)
<b>Total (row %)</b>	<b>12 (2)</b>	<b>30 (6)</b>	<b>72 (13)</b>	<b>96 (18)</b>	<b>108 (20)</b>	<b>137 (25)</b>	<b>65 (12)</b>	<b>23 (4)</b>	<b>543 (100)</b>

## RESULTS

Figure 3: Cause of Death among Californians with SCD 2004-2015, n = 543

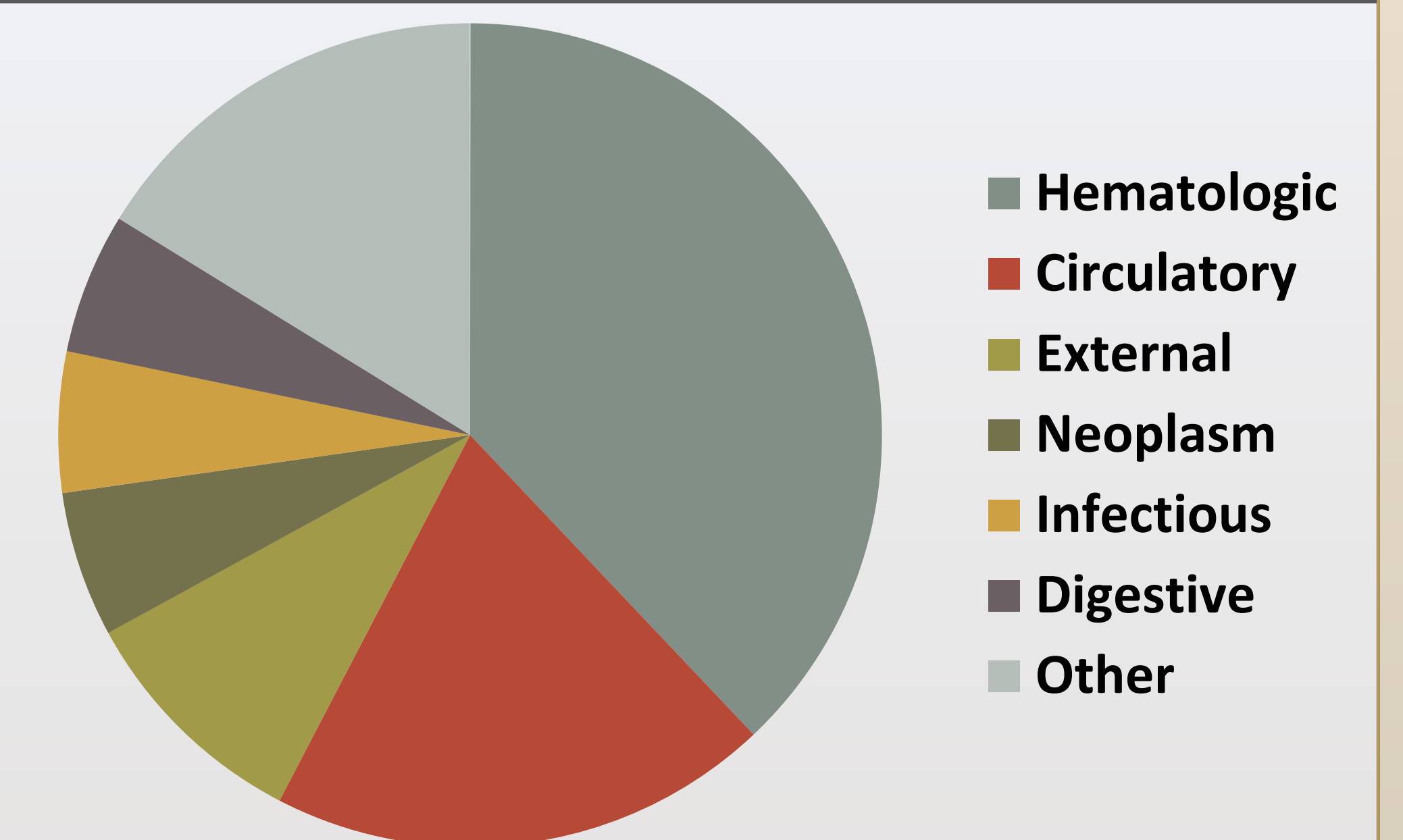
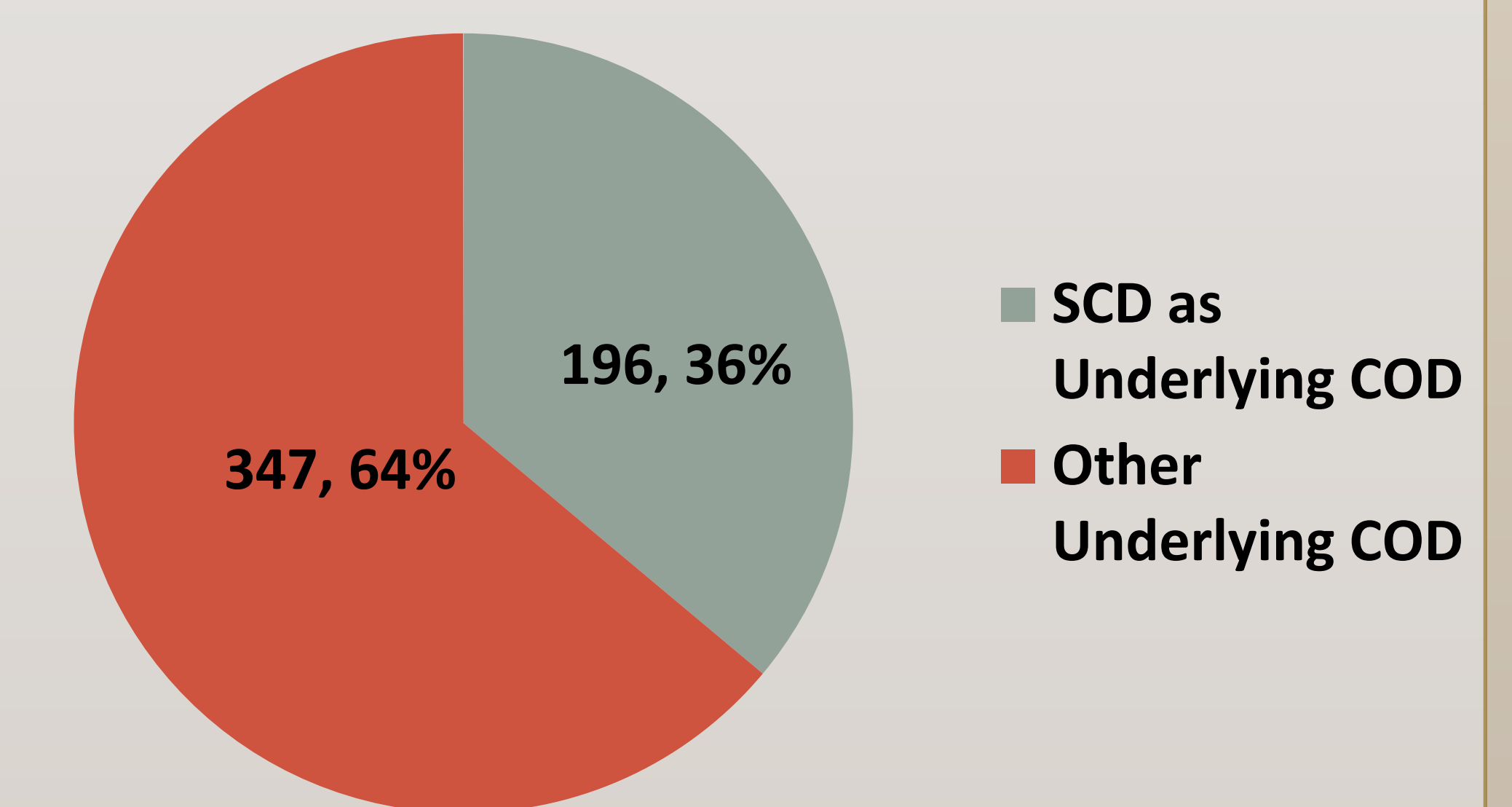


Figure 4: SCD vs. Not SCD as Underlying Cause of Death among Californians with SCD 2004-2015, n = 543



## CONCLUSIONS

- Mean and median age of death are slightly higher than earlier reports based on same methodology (Paulukonis et al, 2016): mean 42.2 years, median 42.8.
- Death among those age 50-59 was most common.
- Death in a hospital setting (inpatient or ED) occurred most often compared to other settings.
- Patients with SCD died from a variety of stated cause of death.
- SCD is not reliably coded in death certificates (36%), making mortality and associated conditions challenging to monitor at a population level.
- Multi-source data collection allows for population based surveillance of health outcomes for those with SCD.

## ACKNOWLEDGMENTS AND DISCLOSURES

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