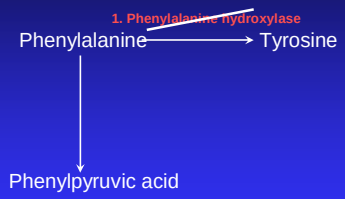
Biocultural Case Studies

Phenylketonuria, Cretinism and Geophagy

Diet Coke

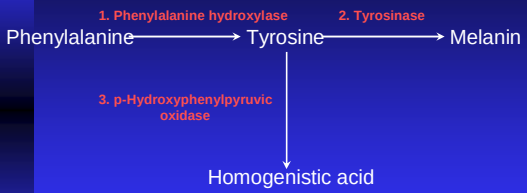
Phenylalanine pathways



PKU

- Phenylalanine hydroxylase insufficiency causes phyenlketonuria (PKU)
  - Most common genetic abnormality in the U.S. (1:10,000 overall, about 1:2,500 Europeans)
  - Growth retardation, mental retardation, depigmentation of skin, hair
  - Screened for in infancy and treated by a diet restricted in phenylalanine intake which causes some growth retardation
    - Different screening techniques have different false positive results
      - About 2/3 of positives have classic PKU
      - New test released in 1998 cuts false positives from about 1% to about 0.01%

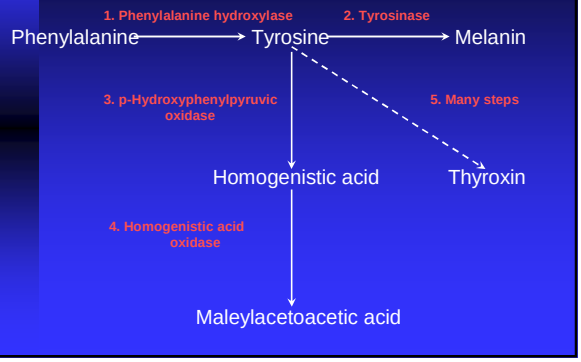
Phenylalanine pathways



Phenylalanine pathways

2. Tyrosinase deficiency causes albinism
   - Can have reproductive implications including enhanced or reduced fertility depending on mating preferences.
3. P-hydroxyphenylpyruvic oxidase deficiency causes tyrosinosis
   - Increased levels of circulating tyrosine and tyrosine metabolites

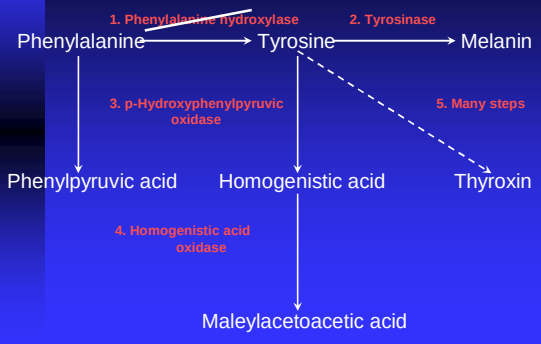
Phenylalanine pathways



Phenylalanine pathways

4. Homogenistic acid oxidase deficiency causes alkaptonuria
   - Black urine, deposits in joints like arthritis
5. Deficient enzyme(s) on this pathway causes hereditary cretinism
   - Similar to iodine deficiency disease, but not treatable by diet

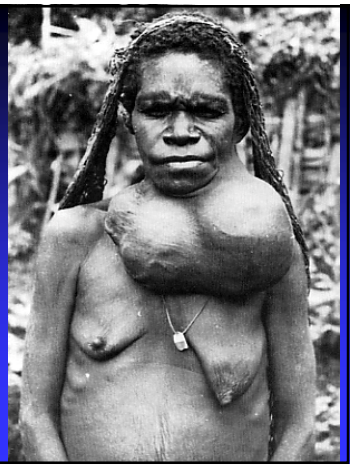
Phenylalanine pathways



Goiter and Cretinism

- Iodine is a necessary constituent of Thyroxin made in the thyroid gland at the base of the neck by combining iodine with tyrosine
  - Thyroxin functions to increase the cellular rate of carbohydrate metabolism and of protein synthesis and breakdown
- Deficiency in production of thyroxin can result in goiter and cretinism
  - Goiter: thyroid hypertrophy
  - Cretinism occurs in children born to goiterous mothers
  - Musculo-skeletal growth retardation, CNS impairment frequently deaf-mute

Dani Woman with large goiter



Goiter and Cretinism

- Endemic pockets:
  - Inland and highland settings, Detroit, Peru
  - Highland New Guinea
- Deficiency can by induced by goitrin present in Cruciferae (e.g., cabbage, rutabagas)
  - Hypothesized relationship with PTC tasting since PTC tasters are sensitive phenylthiourea which is present in most crucifers
  - Thiocyanate contained in yams and cassava also suppresses absorption iodine

Dani Women with Goiters

Cretinism among the Maring

- No goiters noticed prior to 1966
  - 1968 survey found 25% goiter rate among adult females and 24 cretins in a population of about 2,000
    - All cretins less than 8 years old
  - Focus on change in 1960—non-iodized trade salt introduced
    - Traditional salt source contained iodine but took a lot of work
  - Injected iodized oil and required all salt carried into the area to be iodized after 1970

Dani Men with Cretin

Next

Geophagy

Based on a presentation by
Erica Gibson-Staneland

What is Geophagia?

- It is a subcategory of Pica
  - Eating non-food items
  - Pica is derived from the Latin for magpie, a bird with a catholic appetite
- Geophagia is the practice of eating dirt or earthen clays
  - Literally geo “earth” phagia “eating”
  - Clay, laundry starch, and ice are most commonly craved items

Hypotheses

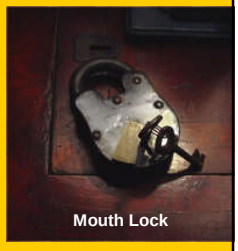
- Pica/Geophagy is practiced for many reasons including:
  - Response to obtain needed nutrients
    - Calcium, magnesium, etc.
      - Virunga Gorillas are known to eat volcanic soils high in potassium
  - Response to hunger
    - Where available, clay is free and filling
  - Cultural phenomenon
    - Learned practice transmitted primarily from mothers to daughters

Hypotheses

  - Response to physiological changes, especially associated with pregnancy
    - Dry up salivary secretions, reduce nausea of morning sickness
  - Way of seeking attention
    - Pregnant women use Pica to seek social support
  - Protect body from toxins
    - Clay minerals bind with plan toxins like glycoalkaloids rendering them harmless

Dirt Eating Around The World

- Geophagy has been found around the world. It appears to have originated in the tropics of Africa, and is now found on every continent except Antarctica
- Slaves brought the custom of dirt eating to the New World with them, although there is some evidence of geophagy in the New World before European contact
  - Aboriginal peoples of the New World mixed earth with potatoes or acorns to neutralize glycoalkaloids or tannic acids so that the food would be easily digestible
- Slave owners were appalled by the dirt eating practices of the slaves, and they put mouth locks on them to prevent them from eating dirt
- The slaves tried to commit suicide by eating dirt so that their bodies would return to their homeland



Mouth Lock

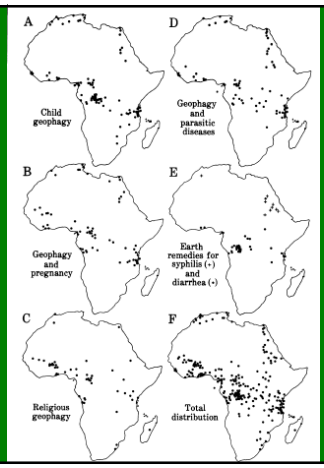


FIG. 2—Processing near a clay-excavation site in Accra, Ghana.

FIG. 3—Retail stall of Yoruba woman trader in Kumasi market, Ghana.

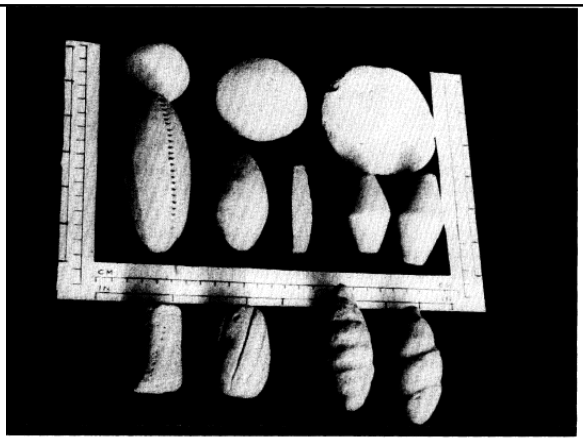


FIG. 4—Specimen geophagical clays collected by the writer in Ghanaian markets.

Iron and Dirt

- Question: Does geophagy cause iron depletion, or is iron extracted from the soil to supplement the women's low iron levels
  - 52 women from a pre-natal clinic in Kenya
  - 73% of the women practiced geophagy, and the median amount of soil consumed was 41.5 g per day
  - No significant link to iron deficiency
  - But locally, geophagy is thought of as a normal behavior for pregnant women

Clay and Pregnancy in Nigeria

- The Tiv are one of the few groups in the world where men also practice geophagy
- The men eat clay as an anti-diarrheal medication
- Tiv women are universal geophagists during their pregnancies
  - They eat clay throughout their pregnancies
    - To alleviate morning sickness in the first trimester
    - Then as a nutritional supplement as the pregnancy progresses
  - The Tiv are a non-dairying population, and women may eat the clay for extra calcium

Geophagy Across Africa

- Wiley and Katz compared geophagy rates among populations in Africa practicing dairying and those who do not
  - Sixty populations were used
  - Geophagy is more common in non-dairying groups except when geophagy is rarely practiced
  - In the six dairying groups that practiced geophagy, three limited women's access to milk

Geophagy During Pregnancy and Dairying in 60 African Populations

	Dairying	Non-dairying
Geophagy rating:		
Absent	5	4
Rare	4	3
Occasional	2	4
Common	2	26
Virtually Universal	2	8

(Wiley and Katz, 1998: 535-536)

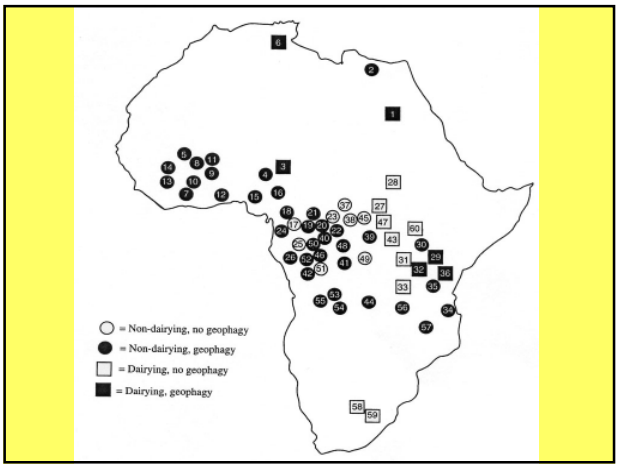


TABLE 5

Clay vs. Dairy Products: Evidence for Similar Activity[1]

Clay	Milk
Calcium for fetal development	Calcium for fetal development
Calcium absorption	Mechanism to absorb calcium (via lactose in milk)
Decreased toxicity of plants	Decreased reliance on plants in diet
Reduced toxins to embryo	Nontoxic to embryo
Recommended for pregnant women in some cultural contexts	Recommended for pregnant women in some cultural contexts
Moisture removed; transportable in loaves; readily available	Moisture removed; transportable as cheese; readily available
Available in tropical Africa	Not available in tropical Africa
Not available in deserts of Africa	Available in deserts of Africa

TABLE II—SIMULATED HUMAN DIGESTION OF CLAY SAMPLES*

(*In parts per million [micrograms per gram]*)

SAMPLE NUMBER	MACRONUTRIENTS				MICRONUTRIENTS			
	Phosphorus	Potassium	Calcium	Magnesium	Copper	Zinc	Manganese	Iron
1	27	108	1,269	298	7	25	0	75
2	7	98	1,400	548	2	30	0	48
3	8	210	190	333	4	18	0	53
4	30	50	1,133	368	10	13	14	393
5	4	150	162	17	13	13	0	47
6	20	200	115	17	4	11	0	40
7	8	130	428	183	11	18	1	58
8	112	180	1,449	386	22	15	0	126
9	60	118	1,229	331	10	23	1	173
10	14	250	946	268	17	11	0	95
11	12	60	1,516	405	8	12	0	262
12	23	80	764	265	9	13	0	175
Median	20	130	1,133	331	10	15	0	95

* The procedure used was 0.5 grams of the sample to be digested with 50 milliliters of 0.1 N hydrochloric acid for six hours by shaking on a rotating shaker at 200 revolutions per minute. The samples were filtered, and the analysis was carried out by the same procedures used for total analysis in Table I. Analysis by courtesy of Dr. Boyd G. Ellis.

Mineral Supplements?

- The Garifuna women of Belize eat clay during pregnancy to alleviate nausea and to satisfy cravings
- Their clay comes in the form of religious tablets that are manufactured in Guatemala
- Eating one tablet per day provides women with **9%** of the RDA of calcium and iron, **7%** of her magnesium, and **3%** of potassium and zinc
- 6 of 8 women interviewed ate more than one bar per day
- This form of supplementation is comparable to Western pharmaceutical supplements "One a Days"

Anemia and Clay Eating

- 40 of 152 women in Mississippi ate clay during their pregnancy
  - about 50 g/day
  - No impact on hematocrits
  - Conclusion: Women in Mississippi practiced geophagy as a result of cultural transmission retained from Africa, rather than from nutritional need
- A second study found that 94% of women geophagists in Mississippi had an inadequate diet
  - Response to hunger?

A woman buying dirt at a convenience store in Alabama.

Summary

- Geophagy may be an adaptive practice, used by women whose physiological need for nutrients drives them to seek out new substances, especially during pregnancy
- Women eating clays during pregnancy may do so for many reasons:
  - to prevent morning sickness
  - provide nutrients
  - detoxify substances for the mother or fetus
  - satisfy a psychological craving which imparts a sense of comfort

Now, the question of the day:

Would you eat dirt or clay?

You probably already have!

There's a Fungus Among Us

Mycotoxins

Mycotoxins in Food

- **Certain molds produce powerful toxins that poison food. A wide range of diseases result from acute mycotoxicosis**
- **Long-term effects include reduced fertility, loss of immune system function, cancer and growth suppression**

Mycotoxins

Certain molds produce powerful toxins that poison food. Farm animals are the most affected today


Fusarium spores

- **Aflatoxins –** acutely toxic and cause liver and esophageal cancer, From *Aspergillus flavus.*
- **Tricothecenes** –produced by *Fusarium* infected cereals
  - Causes alimentary toxic aleikiia (ATA), yellow rain, hole in the head disease
- **Penitremes –** produced by *Penicillium* species
  - Causes Ijesha shakes in Nigerians and grass staggers in sheep.

How common is this?

- Prior to agriculture cereals were small part of diet
  - Fungal toxicity would have been extremely rare
- Since cereal crop domestication, appears to be regular occurrence
  - ATA epidemic in the USSR estimated to have killed approximately100,000 people between 1942 and 1948

Recent Outbreaks

- Aflatoxin in Dog Food caused an epidemic just last year
- 2004 saw an Aflatoxin outbreak in Kenya
  - As of July 20, 2004 a total of 317 cases had been reported with 125 deaths
  - 182 (53.2%) of 342 samples of maize purchased in agricultural markets had >20 ppb of aflatoxin

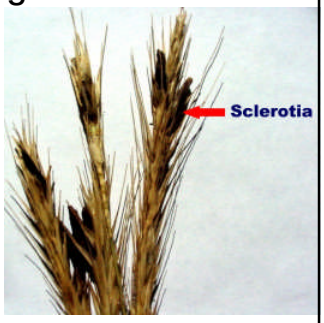
FIGURE 1. Districts affected by aflatoxicosis outbreak — Eastern and Central Provinces, Kenya, January–July 2004

FIGURE 3. A posho (maize flour) mill — Makueni district, Eastern Province, Kenya, 2004

Photo/CDC

Ergot

- The ergot fungus caused by species of *Claviceps* (typically *Claviceps purpurea*), that appears as an infestation on the flowers of many of our cereal crops (wheat, barley, oats, and especially rye), and whose toxic substances (mycotoxins) can be passed right on through the milling and baking process to the final bread product



Ergot

- Medicinal Uses
  - Migraines (ergotamine tartrate)
  - To control bleeding during childbirth (ergometrine)
  - For certain psychiatric conditions
  - To induce labor (since the 1700s)
- Ergotism is poisoning by ergot consumption
  - Symptoms include hallucinations (from lysergic acid hydroxyethylamide, precursor to LSD)
  - Formication (the feeling of being covered by ants)
  - Intense pain in extremities due to constriction of blood vessels

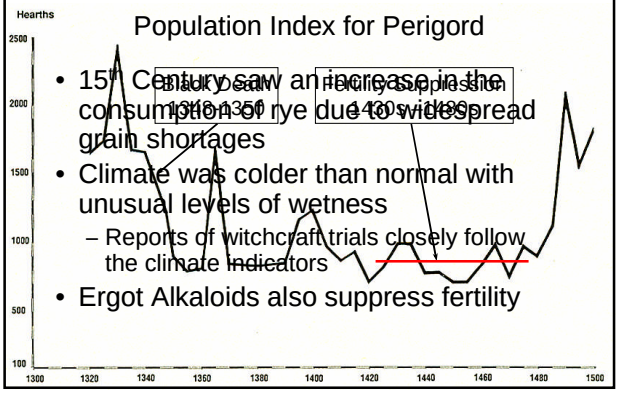
Ergotism

- Mortality can be high (>40%), and victims who survive experience reduced immune response, blindness, and loss of fertility
- Ergotamine is a strong vasoconstrictor, so powerful as to completely block the flow of blood to parts of the body, creating the condition known as gangrenous ergotism
  - Limbs are particularly susceptible and in its initial stages the extremities can feel as though they are on fire
    - This 'plague of fire' or 'holy fire' as it was sometimes called, could set off whole afflicted communities in involuntary dancing movements—the infamous "St. Vitus's Dance" or "St. Anthony's Fire" of the Middle Ages

Ergotism

- Historically significant as a cause of widespread panic, population suppression, and extreme religious behavior
  - Demographic depression 1430s – 1480s
  - The Salem Witch Trials
  - The Panic of 1789

Reduced Fertility



- 15th Century saw an increase in the consumption of rye due to widespread grain shortages
- Climate was colder than normal with unusual levels of wetness
  - Reports of witchcraft trials closely follow the climate indicators
- Ergot Alkaloids also suppress fertility

Salem Witchcraft

- Rye was a common grain growing in the area before the Puritans arrived
  - There was dissatisfaction with the rye because it made the animals sick
  - Climate in 1691 was conducive to ergot
    - Early rains and warm weather in the spring progressed to a hot and wet summer
    - 1692 was a drought year, dramatically reducing ergot
  - Threshing probably took place shortly before Thanksgiving
    - Children started showing symptoms in December
    - By late fall 1692 the crisis abruptly ended

Salem Witchcraft

- Rye growing in low, wet ground is most susceptible to ergot growth
  - One of the most notorious of the accusing children in Salem was Thomas Putnam's 12-year-old daughter, Ann
  - Her mother also showed symptoms
  - Two other afflicted girls also lived in the Putnam residence
  - Putnam had inherited one of the largest landholdings in the village
    - His father's will indicates that a large measure of the land, which was located in the western sector of Salem Village, consisted of swampy meadows

Salem Witchcraft

- Much of the western acreage of Salem Village may have been an area of contamination
  - This is substantiated by the pattern of residence of the accusers, the accused, and the defenders of the accused
  - Excluding the afflicted girls, 30 of 32 adult accusers lived in the western section
  - 12 of the 14 accused witches and 24 of the 29 defenders lived in the eastern section
- The general pattern of residence, in combination with the well-documented factionalism of the eastern and western sectors, contributed to the progress of the witchcraft crisis

Parris: Two afflicted girls, the daughter and niece of Samuel Parris, lived in the parsonage almost exactly in the center of the village. Their exposure to contaminated grain from western land is also explicable. Two-thirds of Parris's salary was paid in provisions; the villagers were taxed proportionately to their land-holding. Since Putnam was one of the largest landholders and an avid supporter of Parris in the minister's community disagreements, an ample store of ergotized grain would be anticipated in Parris's larder. Putnam was also Parris's closest neighbor with afflicted children in residence

person, was accused of witchcraft by the other afflicted girls, and then became afflicted again. Two depositions filed against her strongly suggest, however, that at least her first affliction may have been a consequence of ergot poisoning.

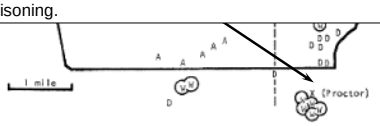
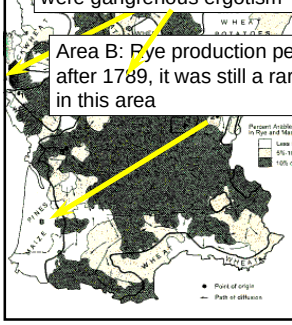


Fig. 1. Residence patterns, Salem Village, 1692. The names in parentheses indicate the households in which the afflicted girls were living, excluding Sarah Churchill, whose affliction is believed to have been fraudulent. The nonvillagers shown on this map are those whose places of residence lay on the fringes of the Village boundaries. Residences are labeled (X), afflicted girl; (W), accused witch; (D), defender of the accused; and (A), accuser. [Adapted from Boyer and Nissenbaum (*Id.* 75)]

Panic of 1789

Area C: Rye carefully cleaned because of earlier episodes of St. Vitus' dance—most effects here were gangrenous ergotism

Area B: Rye production peaked after 1789, it was still a rare crop in this area



- July 20 – August 6 waves of panic swept the French countryside
  - Coincide with the start of the French Revolution
- Rumors that the rye crop was to be seized
- Medical reports consistent with ergotism
- At least one eye-witness account of prodigious ergot infection of the rye crop
- Most favorable climate for ergot in France for almost 100 years