PERSISTENT, ABERRANT MONGOLIAN SPOTS

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THIS paper is devoted to the subject of persistent, aberrant mongolian spots, and a few comments will be made on differentiation of this condition from extensive blue nevus. Four cases are reported.

REPORT OF CASES

Case 1.—Interest in the subject was first aroused through observation of a 23 year old Negro woman who presented a pigmented area on the left side of her face and nose, and bluish spots in her left eye, which spots had been present since birth. There was a palm-sized macular area of grayish green pigmentation on the left side of her face, extending from in front of the ear to the angle of the mouth, with a smaller spot on the side of the nose. In addition there were four discrete bluish macules on the left sclera.

Histologic examination revealed a normal epidermis and dermis except for hemosiderin-free, dopa-positive melanoblasts situated in the middle third of the corium. The coarse pigment granules were contained in stellate and spindle-shaped cells which were quite dispersed and not sharply circumscribed in clumps. The upper portion of the corium was free of pigment and nevus cells. (Slides were later submitted to Dr. Hamilton Montgomery, who concurred in the diagnosis.)

This patient was presented before the Cleveland Dermatological Society as having an aberrant, persistent, mongolian spot.¹

Case 2.—Through the courtesy of Dr. Hamilton Montgomery and by consent of the authors, we are able to incorporate in this report material in connection with the case of Jensen and Haffly.²

Under the title, "An Unusual Case of Progressive Melanosis Oculi," Jensen and Haffly described the case of a 40 year old Chinese with a pigmentary disorder involving the right eye, the right side of the face, the forehead and the scalp. According to the patient, his parents had first noted a small pigmented

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Fig. 1.—Extensive grayish green spots on the cheek and the sclera in a Negro woman aged 23 (case 1).

Fig. 2 (case 2).—Persistent mongolian spots of the right side of the face and the conjunctiva bulbi (courtesy of American Journal of Ophthalmology and Drs. Jensen and Haffty, Seattle).
area on the right eyeball when he was 5 years of age. During the first two decades the lesion gradually increased in size but remained confined to the globe. In the second two decades, the pigmentation spread extensively over the right side of the face, nose, forehead and scalp.

As the authors put it, "There was a diffuse slate-gray, non-elevated pigmentation irregularly spread over the right side of his face, extending through the floor of the nose into the mucous membrane of the mouth. In the mouth the pigmentation was confined to the hard palate. The overall pigment picture might be likened to the end result of a severe powder-blast burn." The conjunctiva, sclera, cornea and iris of the right eye and the iris of the left eye were similarly involved.

Fig. 3 (case 3).—Sixty-three year old white man with mongolian spot on the upper part of the back and the neck.

In the histologic sections, throughout the corium were numerous cells varying from stellate to fusiform in shape, filled with brown pigment. The cells were dispersed and not densely clumped, as might be expected in a blue nevus. There were no nevus cells. Unfortunately, no studies on the dopa reaction were made at the time, and it is not possible to make the test at present.

Case 3.—Also through the courtesy of Dr. Montgomery, the material from another case, originally seen by Drs. Montgomery and Brunsting at the Mayo Clinic, has been made available to us.

The patient was a 61 year old white man who presented an irregular, flat, slate gray plaque, measuring 17 cm. in diameter, over the upper portion of the back and neck. The lesion had been present since early childhood. In the report of the microscopic examination, Montgomery stated, "The histologic picture is
that of a mongolian spot plus senile skin changes. Pigment bearing cells in the cutis are spaced much farther apart than one sees in the ordinary blue nevus.” The dopa reaction was positive.

CASE 4.—The final case is that of a 25 year old, blonde white woman. The patient stated that, according to her parents, a bluish discoloration was noted on the eyes at birth. This same process was first observed on the eyelids in very early childhood, and while there was some progression and increase in intensity during the first decade, the condition remained localized in the eyes and on the lids until early adolescence. During the second decade, there was extension of the condition onto the forehead and temples. It was felt by the patient that since the twenty-first year there had been definite regression. We had, at the time of writing, been following this patient for over a year, and it was our impression, also, that the intensity of pigmentation was fading.
Fig. 6 (case 4).—The dopa reaction of the skin of the forehead was weakly positive; $\times 354$. Note the scattered, fusiform cells with processes in the mid-corium.
The family history was not significant. There were twelve siblings: seven brothers and five sisters. The patient was of pure Anglo-Saxon and Irish stock. No other members of the family were similarly affected. There was no evidence on physical examination, or from the history, of consanguinity, neurofibromatosis (von Recklinghausen's disease) or other pigmented disorders.

Clinical examination revealed numerous discrete bluish spots on the bulbar conjunctivas of both eyes. The lids of both eyes showed a deep, macular, diffuse grayish blue discoloration, and as the process extended upward onto the forehead and temples, it became less and less evident. The almost perfect symmetry was an additional striking feature. At the periphery of the lesion, the pigmentation appeared to fade away gradually and blend into the color of the normal surrounding skin.

The patient insisted that the specimen for biopsy be removed from a very light blue area next to the hair border. Permission for the taking of further specimens was denied.

The sole feature presented by the microscopic section was the pigmentary abnormality confined to the dermis. There were numerous, scattered fusiform cells containing pigment granules which gave the histochemical reaction for melanin as demonstrated by silver impregnation. These cells were most pronounced in the midcutis and lower portion of the corium. The dopa reaction was weakly positive.

HISTORICAL REPORTS

Meirowsky stated that the Danish missionary Soabye in 1778 reported that the children of Greenland were born even as white as the Europeans but that they laid a 10 shilling piece-sized bluish-mark over their sacral area which gradually spread over the entire body and was, perhaps, the cause of their dark complexion. Another Danish missionary, Hans Fabie in 1816 mentioned the presence of a bluish birthmark on the sacrum of the children of Greenlanders, and Eschricht in 1849 described an analogous process in Eskimos. Riedel, as early as 1847, noted the so-called mongolian spots on babies in the Celebes and wrote about them to Charles Darwin. Darwin expressed the belief that the spots were an atavistic indication that the forebears of the patient had had a tinted back side.

NAMES FOR MONGOLIAN SPOTS

In 1905 an American physician, A. S. Ashmead, for a long time head of a large hospital in Japan, wrote of the spot as "the mulberry colored spot." He stated that it was called in various localities as fol-


lows: Samoa, ole-ila; Hawaii, he-ila; Central America, nits gran; Negroid sections of Brazil, geni papo; coastal province of Argentina, mancha morata, and Germany, Mongolen-fleck. In France it is variously designated as tache bleue, tache mongolique, tache pigmentaire and tache bleue sacrée.

**INCIDENCE OF THE LESION**

As would be expected, the incidence of this spot varies with different races and parts of the world.

El Bahrawy asserted that the spot or spots will be found in 90 to 100 per cent of Mongolians, 100 per cent of Malayans, 85 per cent of Bolivian Indians, 65 per cent of Negroes in Brazil, 52 per cent of mixed races in Brazil, 2 to 5 per cent of Italians in Brazil, and 1.5 per cent of white persons in Brazil. One per cent of white persons in Paris will have the spot, and 7.5 per cent in Montevideo, he stated. Von Bulow, quoted by Meirowsky, said that on persons born from the union of white persons and Samoans the spot is not so frequent but that on those from the union of halfbreed Samoans and Samoans they are very common.

Fischer and Shen Chen Yü estimated that 90 per cent of Polynesians have mongolian spots and 90 to 99 per cent of Japanese. They expressed the opinion that 98 per cent of the newborn in China have the spot, and 90 per cent of the newborn in Japan.

Chemin studied the frequency of occurrence of the spots in China and found them in 89 per cent of children 0 to 1 year old, 71 per cent of children 1 to 3 years old and 19 per cent of children 3 to 8 years old.

Adachi stressed the point that it is only a difference in percentage between the incidences in Mongolian and in European stock. As El Bahrawy well put it, “The mongolian spot is no exclusive index of the Mongolian races, but rather a well spread manifestation of the entire human race.”

**APPEARANCE, DISAPPEARANCE AND PERSISTENCY OF THE LESIONS**

The mongolian spot is generally present at birth or shortly thereafter, rarely later. In a large proportion of cases it will gradually fade out over a period of a few years; witness the previously cited figures

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of Chemin's findings in China. Katō⁹ made the assertion that the pigmentation gradually fades up to 4 years and thereafter more rapidly, until by the thirteenth year it is seldom seen.

Fischer and Shen Chen Yü,⁶ working in Shanghai, found it in 57 of 63 patients under 4 years of age, in 23 of 47 patients over 4 years and under 7 years of age, in 3 of 15 patients over 7 years and once in a 14 year old child. The spot, they stated, rarely lasts the entire life. Nevertheless, it may occur even if it does not show grossly. Ishikawa,¹⁰ in histologic studies of the bodies of Japanese from the ages of 18 to 82, found typical "mongolian cells" in all of them although the cells were less frequently encountered in examination of older persons. Moreover, study of the fetus revealed the gross appearance of the spot by the fifth to sixth month, and Ishikawa found the pigment cells in the beginning of the third fetal month in very sparse numbers. From the fifth month on, they were present almost without exception.

SITE OF THE LESIONS

The typical site of the lesion is well known to all—the region of the lumbosacral junction and perhaps the contiguous inner aspect of the buttocks. In a fair proportion of the cases that area is the only site. El Bahrawy said that far less frequently the lesion is found on other parts of the body—breasts, abdomen, extremities and face. It may be present at birth or appear somewhat later. In adult Japanese it is seldom found and then in relatively unusual locations. El Bahrawy quoted Balz to the effect that, rather than the spots being a typical identification mark of the Oriental, analogous spots may be found in all races and peoples, although in different incidences and intensities.

Wateff¹¹ studied the frequency of occurrence of the lesion in Bulgaria and said that, in addition to the areas stated, it will be found rarely on the back and very rarely in other areas. Fischer said that spots on the ventral surfaces are much rarer. He stated that Cozzolino saw a male child with the typical sacral involvement and another spot on the lower part of the thigh, whose brother had a 7.5 by 5.5 cm. spot on the left side of the anterior portion of the pelvis. Wateff thought there was more tendency to involvement of the left side of the body among the 3,500 infants (Bulgarian, Macedonian, Jewish and Bohemian) that he studied and among whom he found 20 bearers of one or

more spots. All had the sacral involvement. Several had bizarre, diffusely pigmented areas of the trunk—in 6 of them, over almost the entire back. One had a spot on the dimple of the upper lip and another lesion on the chin, while 1 patient had two spots on the vertex of the head.

In Ebert's case,\(^{12}\) of a Mexican child aged 2½ months, there were bluish black macules on the buttocks and sacrum, several coin-sized bluish spots over the back, a large spot on each shoulder and a lesion on the left knee.

In discussion of Ebert's case, Drs. Edward Oliver Sr. and Clark Finnerud stated that they had observed a patient a year earlier with spots on the leg, several on the body, a well demarcated one above the ankle and one on the thigh. Eleven months later these spots were much paler. Dr. Finnerud stated that he had also seen an Indian infant with almost half of the body, including the face, neck, ears and lower extremities, involved.

Adachi observed spots situated in front and in back of the ears. Sekiba\(^ {13}\) observed the mongolian spot on the face twice in adults, and said that a colleague, Tashiro, had a child of his own with many extensive spots on the buttocks and extremities, and that the father removed one from the forehead surgically. Königstein, according to Wateff,\(^ {11}\) observed the condition on the back of the hand and on the neck.

Strangely enough, involvement of the region of the eye is not unheard of. Adachi,\(^ {9}\) in his extensive study, pointed out that in anthropoid apes there is a strongly pigmented skin of the eyelids even in those that have a relatively pigment-free skin on most parts of the body. The conjunctivas of apes, he stated, is not pigment free.

Reese,\(^ {14}\) a well known American ophthalmologist much interested in the problem, made the assertion, "The extrasacral spot and the blue nevus are seen principally on the face and extremities, and may appear on the lids and bulbar conjunctiva."

Another well known American physician, William Allen Pusey,\(^ {15}\) as long ago as 1916 reported the case of a Chinese university student with a flat, brown to black nevus, not hairy, of the right side of the face. There was, in addition, a spot of bluish pigment on the sclera.


Leber and Prowazek\textsuperscript{16} stated that there is occasionally a pigmentation of the scleras in the natives of Samoa and that it gradually fades with age.

Fischer and Shen Chen Yü observed the spot on the bulbar conjunctiva three times and have seen spots on the body the size of the flat of the hand.

COLOR OF THE MONGOLIAN SPOT

The color of the mongolian spot has been variously described as light blue, blue, grayish blue, dark blue and mulberry. It has been compared to that of the tattooage observed from gunpowder. Occasionally it may be a slate gray tint. At times there may be a main island with lighter spots around it. In Negroides the lesion may even be grayish green, as observed in our case 1, and ordinarily the tint is darker in a person with a dark complexion. In Eurasians the color is lighter and generally disappears earlier.

SIZE AND SHAPE OF THE LESIONS

The lesion may be irregularly round, roughly triangular, ovoid, heart shaped, angular or formed like a horseshoe, lance or tennis racquet. Only very exceptionally there may be hairs. Meirowsky\textsuperscript{8} and other authors emphasized the absence of hair. The area may vary in size from 0.5 to 5.0 cm. in diameter, and Bruch\textsuperscript{17} said that the spot may extend over a very large area. Fischer and Shen Chen Yü\textsuperscript{6} stated that the size ranged “from that of a bean to the flat of the hand.” The entire half of the back may be included. Zebra-like configurations may be found.

MICROSCOPIC OBSERVATIONS

The characteristic picture in this condition is the finding of the so-called “Mongolian cell.”

El Bahrawy,\textsuperscript{4} an authority on the subject who studied at Bloch’s clinic, described the cells as long, spindle shaped, irregularly wavy, 5 to 10 microns thick and 30 to 50 or 100 microns in length. The center is the widest portion with a pale, oval, lightly staining nucleus that shows up in an unstained specimen as a light spot. The cells may have long processes and may be bipolar or irregular. Some cells are flat, others pear shaped.

Grimm\textsuperscript{18} said that the processes of the cells may be serpentine or straight, stretching out into the connective tissue, even seemingly to

\begin{footnotes}
\item[17.] Bruch, A.: Notes sur la tache bleue sacrée dite mongolique, Arch. de méd. d. enf. 15:446, 1912.
\end{footnotes}
anastomose with neighboring cells. In children of the white race, the pigment granules may be so few that the cells are difficult to differentiate, especially in older children.

Adachi, a Japanese authority on the condition, stated that the cells are found deep in the corium of the pigmented areas. Katô said that in the first week of life they may be found even in the upper part of the corium as well—later, only in the lower portion. They are arranged between the connective tissue fibers and usually lie parallel to the epidermis. Some may be found around the vessels.

In fresh unstained material from 10 cases in the newborn at Shanghai, Fischer described the pigment as light yellow or brown, or a much darker, yellowish brown, brown-black or black. The pigment in fine kernels fills the entire cell.

Adachi stated that in apes, while there is some variation, the cell is found almost uniformly in the deep part of the corium. He pointed out that there are two kinds of pigment cells in the corium of man and apes: one small and in the upper layers, and another, deeper, the deeply placed cell being the mongolian cell. All the authors stress the location of the cells in the lower third of the corium.

They begin to appear in the foetus as early as the third month, and from the fifth month on they are present almost without exception. . . . Moreover, even after the disappearance of the spot anatomically, microscopic study in corpses may reveal their presence from thirteen to eighty-two years.

Masson and Becker differentiated the pigment cells in the upper part of the corium as melanophores (carriers of pigment), the more deeply situated cells being classified as pigment-producing melanoblasts. The latter are "dopa positive." Masson appeared to look on these cells in a way as glandular cells; i.e., producers of pigment. However, he stated, "Stress must be put on the fact that a melanoblast may lose, at the same time, its dopa reaction and its aptness to produce pigment." It is our feeling that 1 patient whose case is reported here (case 4) is probably in this stage. Moreover, she apparently is losing some of the intensity of her pigmentation. Bloch himself presented 2 babies illustrating this point before the Swiss Dermatological Congress at Zurich in July, 1920. The slate-colored spots on the sacrum of each showed cells with melanin, black on contact with silver nitrate and osmic acid and light on

contact with hydrogen peroxide. For both the dopa test was negative. Masson would put it thus in such a case: "The melanoblast persists as a cell and deserves to retain its name, but it may be considered as inactive."

Montgomery and Kahler said that the melanoblasts in the blue nevus are denser and may be grouped in irregular masses in the lower two thirds of the cutis, but that they may even be found high up under the epidermis. They are thickest in the center and give a positive dopa reaction, and disturb the normal arrangement of the connective and elastic tissue. Moreover, the blue nevus is usually raised slightly above the surface. It may be localized or rarely diffuse. Thus, Upshaw, Ghormley and Montgomery reported the case of a white boy, aged 9, with an extensive blue nevus on the left flank, 17 cm. long and 3 to 6 cm. wide. Microscopic study showed the cells of the growth to be dermal melanoblasts, while nevus cells, pigmented in type, were not present. The melanoblasts had long, bipolar, dendritic processes and the dopa test was positive. The dense masses of cells as seen in the lower dermis are never seen in the mongolian spot. Moreover, judging from clinical experience and a study of the literature, we are not aware that any true mongolian spot shows the "multiple, fine, dark blue, round nodules, slightly raised above the skin and with the bluish discoloration in the intervening small areas between the circumscribed nodules." These two items clearly differentiate the mongolian spot and the rare extensive blue nevus.

**COMMENT**

In this connection, Carleton very recently presented a remarkable case of bluish, bruiselike spots of pigmentation in a white girl aged 14. It appeared first at birth on the sclera and iris of the left eye, and later, at the age of 3, on the back, behind the ears, margin of the forehead, nose, lips and vestibule of the mouth. Some of the spots kept forming up to the age of 9 years, and once formed they did not alter their appearance. The girl also had a bossy thickening of the skull behind the ears, although there was no evidence of increased intracranial pressure or cerebellar dysfunction. Carleton very expressively described the color of the sclera as similar to the stain from "spilt ink." Microscopic examination of the skin revealed a widespread infiltration of dendritic bipolar cells containing fine brown pigment granules giving the histochemical reaction of

mela nin. The cells were around the blood vessels and sweat ducts, and scattered through the dermis, particularly the deeper portion. One specimen gave a positive, another a negative, reaction to the dopa test.

In discussion, Parkes Weber expressed the opinion that the condition was a "forme fruste" of neurofibromatosis, showing pigmentation only, and he stated that in 1 of his cases the color was likewise blue. The point was raised by Carleton that neurofibromatosis is a justifiable diagnosis only in the event of a strong familial history. In her case there was no such history. There was no consanguinity, and the patient was of pure British stock and the only one affected in a family of three.

Carleton stated that she did not think this condition could be a blue nevus, which is localized, nor a mongolian spot, which is rare in Europeans. In the latter, the lesions are present at birth and gradually disappear. She expressed the opinion that in the distribution and in the appearance of spots on the sclera at birth, becoming more conspicuous up to the age of 9, the disorder differed considerably from mongolian spots.

Carleton stated that she wondered about its possible relation to a diffuse melanosis described by Hadwen in cattle and horses, especially Percherons, which grow gray and white with age. In these, melanin-containing cells are found around sweat glands and blood vessels and also in the periosteum, lymph nodes and muscles. The deposits begin early, at 6 years or more, and gradually increase.

Carleton expressed the opinion that her case represented a diffuse abnormality in mesodermal pigmentation not exactly comparable with any hitherto described condition and associated with an unusual cranial abnormality.

Except for the osseous changes in the skull in Carleton's case, there is much similarity between her case and our case 4. In neither was there a familial history of neurofibromatosis nor of any pigmentary disorder. The involvement of the vestibule of the mouth in Carleton's case and in the case of Jensen and Haffly are likewise quite similar, and as far as we can find they are the only cases in the literature showing this form of involvement. As has been mentioned, different authors emphasize that in aberrant types of mongolian spot and of blue nevus the face and conjunctivas are prone to be involved.

CONCLUSIONS

While the mongolian spot is seen most frequently over the sacral area, it is also encountered on other parts of the body. Reese made the statement, "The extrasacral spot and the blue nevus are seen principally on

the face and extremities and may appear on the lids and bulbar conjunctivae."

The number, size and shape of the spots vary greatly.

The color varies all the way from a light blue in a blonde to a grayish green in the Negro.

It is found most frequently in the Mongolian and Negroid types, but may be encountered in white persons as well, though such an occurrence is relatively rare.

Rarely the mongolian spot or spots may appear after birth and continue to spread for a period of years.

Microscopically the so-called mongolian cell, found in the lower part of the cutis, is a long fusiform cell, 5 to 10 microns wide and 25 to 75 microns long, often having several dendritic processes. The pigment in an unstained specimen is found throughout the cell as very fine granules, light brown to a darker yellowish brown. The pigment is melanin with a positive dopa reaction. Rarely, in a receding spot, may the dopa reaction be negative. The cells are found between the connective and elastic tissue fibers and are parallel to the epidermis. They are not clumped together, causing distortion, as in a blue nevus.

The very rare extensive blue nevus clinically differs greatly from the mongolian spot. The color is not uniform, and the surface of the skin may be irregular.

A report is made of 4 cases of aberrant, persistent mongolian spot; one of the back and neck, and 3 showing extensive involvement of the eyelids, forehead and bulbar conjunctiva. In 1 instance there was also an area on the hard palate. This last case and the probable case of Carleton's are the only 2 cases thus far reported with mongolian spot of the oral surfaces.

Addendum.—Through the courtesy of Dr. Hamilton Montgomery, we have had the opportunity of reviewing a study by Drs. Harry Pariser and Herman Beerman which has just appeared in print, entitled: "Extensive Blue Patchlike Pigmentation: A Morphologic Variant of Blue Nevus? Persistent Extrascral Mongolian Blue Spot? Diffuse Mesodermal Pigmentation?" 26

A white woman aged 34 first noticed a diffuse bluish pigmentation of the forehead at about 20 years of age. This later spread to the scleras of both eyes and to the cheeks. Histologic examination showed a band of melanin-positive, spindle-shaped cells in the midcutis. There was no disruption of the connective tissue or elastic tissue.

While the histologic changes were more suggestive of mongolian spot, Drs. Pariser and Beerman expressed the opinion that this variant might better be termed blue nevus.

It is very difficult, if not impossible, to draw a sharp dividing line between mongolian spot and blue nevus, but for reasons cited in this paper we wonder whether it would not be preferable to attempt to more sharply define and distinguish the two conditions.

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ABSTRACT OF DISCUSSION

Dr. S. R. Mercer, Fort Wayne, Ind.: We are fortunate that the authors have been able to report 4 cases of such an unusual dermal and ocular melanosis. Clinically, the disorder is harmless, except from the cosmetic viewpoint. I have been able to find no record of melanoma originating in such a lesion. Etiologically the appearance of dermal melanosis, in the form of either mongolian spot or blue nevus, is usually thought to be atavistic. Some of the anthropoids normally exhibit dermal and ocular melanoblasts. That the incidence of sacral spots does not parallel the depth of epidermal pigmentation is apparent from the fact that these spots are seen in practically 100 per cent of Mongolians, who are not always deeply pigmented. MacFarlane (Science 95:431, 1942) found sacral spots in 38 of 50 Bengalese infants. One nurse from the Himalayan region stated, "We call it the sign of a Hill baby," the Hill people being Mongoloid, Nepalese and Tibetan. Sacral spots were found by MacFarlane in all of 15 Javanese babies, but not in those of Dutch parentage or hybrids of Dutch and Javanese. Matus (South African M. J. 15:121, 1941) found with regard to the South African Negro that the pigmented spot was present macroscopically in 75 per cent of the infants studied. His material was drawn from a study of a group of 50 female and 50 male babies, the exact number of spots being 35 in the male group and 40 in the female babies.

Brennemann (Arch. Pediat. 24:426, 1907) estimated that the sacral spot occurs in at least 95 per cent of the Negro children in Chicago. It is evident that racial as well as hyperpigmentary factors are involved.

From the academic viewpoint concerning nomenclature, the use of the term "aberrant mongolian spot" may be questioned. One might insist that the term be used only to designate blue plaques seen in association with sacral spots. Such a sacral spot was not mentioned in any of the 4 cases. The true mongolian spot usually fades out during the first few years of life, while the aberrant spots may have their onset later in life and extend to cover larger areas.

Bettley (Brit. J. Dermat. 50:181, 1938) expressed preference for the designation "progressive melanosis" and stated that, clinically and histologically, the plaque which had appeared at the age of 14 in his 30 year old patient resembled a blue nevus. He also suggested a resemblance to benign progressive melanosis as described by Hadwen in horses and cattle. To my mind the latter process is more extensive and deeper; it approaches malignancy. On the other hand, blue nevi are usually smaller than the large plaques described by the authors. The Japanese (Tanino, H.: Jap. J. Dermat. & Urol. 47:51, 1940) called a large bluish plaque on the face, often associated with ocular melanosis, "nevus fusco-caeruleus."
It may possibly be that the acquired progressive blue plaques bear as close a relation to mongolian spots as acquired nevus pigmentosus bears to the prenatal variety.

DR. HERMAN BEERMAN, Philadelphia: The authors have presented in admirable fashion the subject of persistent, aberrant mongolian spots based on the pertinent literature and four representative cases. The last word has not been said as to the exact nosologic position these flat, extensive, blue-black spots occupy. My interest in this subject was occasioned by study of an example of extensive blue patchlike pigmentation in a white woman of 34 years, who was first seen by Dr. Harry Pariser of Norfolk, Va.26 The patient had first noted pigmentation on the right side of the forehead at about the age of 20 years. The process gradually spread until in 1946 there was involvement of the left side of the forehead, both cheeks and the left forearm. Careful study revealed a histologic appearance similar to that in Dr. Cole's cases, with particular concentration of the pigment cells (so-called mongolian cells) in the midcutis and with some tendency for concentration around hair structures. Our case departs from the original clinical description of the blue nevus of Tièche in three ways. First, the lesion is a bluish nonindurated patch involving an extensive area of the forehead, scalp, face and sclera. Second, it did not appear at birth or during childhood. Third, there is an extension of the process with age in contrast to the original description that the lesions remain static. Objectively this bluish pigmentation resembles the clinical description of extrasacral mongolian spot more than blue nevus. It differs in its course from the usual evolution of extrasacral mongolian spot in that it increased in intensity with age and appeared for the first time in adult life.

Because of the strikingly similar histologic picture of blue nevus, mongolian spot and the variant found in my patient, I suggest that all these conditions be included in the concept of a single clinical entity. In order to avoid introduction of any new terminology, the name blue nevus seems most appropriate for this entity, particularly since the spot can occur in the non-Mongolian races and because this term mongolian spot fails to convey any indication of thenevroid origin or clinical appearance of the lesion.

DR. F. A. ELLIS, Baltimore: Dr. Cole has partly answered my question; I was interested in the problem of what happens to a mongolian spot as the patient gets older. It would be interesting to determine the percentage of melanotic babies that have the mongolian spots and at autopsy, the number of adults that still have the melanin-containing cells in the dermis in the buttocks. Why do the spots clinically disappear? I doubt that the melanin cells disappear, but I think that changes occur in the overlying structures so that pigment is no longer visible clinically.

DR. HAMILTON MONTGOMERY, Rochester, Minn.: The picture of the patient with extensive blue nevus which Dr. Cole showed is from a case reported by Drs. Upshaw, Ghormley and myself.23 Histologically, the condition was a blue nevus with dense masses of dendritic cells in the corium. Fundamentally, there is no great difference between the cells of mongolian spot and those of blue nevus, but in the mongolian spot the cells are widely separated. The ordinary blue nevus usually occurs as a solitary lesion, or there may be a couple of lesions in the same patient. The blue nevus may occur anywhere on the body and has been mistaken for melanocysticuloma. It does not look like the aberrant or misplaced mongolian spot which Dr. Cole has presented. I, therefore, would prefer not to simplify the terminology and call everything a blue nevus. I agree that the term "mon-
mongolian spot" may be a misnomer, just as "sarcoid" is a misnomer. I think that these aberrant mongolian spots with cells widely separated, as they have been in the cases seen at the Mayo Clinic, in the cases of Jensen and in the cases of Cole and his associates should be separated from blue nevi.

Malignant change has not been reported in mongolian spot so far as I know. It has been reported in approximately 6 cases of blue nevus, 2 of which were reported by Darier and 1 by Stout in New York. In all the cases there was a history of preceding repeated irritation and trauma. Thus, we have the same story as in ordinary pigmented nevus, whether it is a junction nevus or hairy deep subepidermal mole developing into melanoeplithelioma after repeated irritation and trauma. When a blue nevus undergoes malignant change, it is then a true melanosarcoma because the blue nevus cells are presumably of mesodermal and not of epidermal origin.

Dr. H. N. Cole, Jr., Cleveland: I should like to make the acknowledgment to Dr. Montgomery for the use of a photograph and photomicrograph of a 9 year old white boy whose case was reported by Drs. Upshaw, Ghormley and Montgomery. In reference to Dr. Ellis's question, it is difficult to say what becomes of the mongolian spot. Most commonly, the mongolian spot is present at birth and fades out rapidly over the period of a few years. Various workers have reported that at birth anywhere from 90 to 98 per cent of the members of the Mongolian, Chinese and Japanese races have these spots over the sacral region. Approximately 3 to 8 per cent retain these spots as late as the middle teens. In Jensen and Haffler's case, the patient had the condition confined to the eye and lid up to 20 years of age. It was only after that time that the pigmentation started to spread over the face onto the forehead and scalp. At the time of publication of their article the condition was still progressing.

The condition in our case, which seemed to be quiescent for a long period, apparently is now regressing. In other words, any number of possibilities may occur. I am not aware of malignancy having been reported in connection with mongolian spot. Whether these lesions should be termed mongolian spot of the extrasacral variety or extensive blue nevus is difficult to say. Of course, if there is dense clumping of the melanoblasts which causes distortion of connective tissue and if the lesion is raised above the skin, I think one is justified in calling the disorder an extensive blue nevus. However, in a case in which there is not such dense clumping of the dermal melanoblasts and in which the lesions are not raised, the category to which the case belongs is certainly open to question.